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Fibrous tumours in children: imaging features of a heterogeneous group of disorders

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Abstract *Background*. Fibrous tumours are predominantly soft tissue lesions which are relatively frequent in childhood but are little known. Imaging is often used in the evaluation of these tumours but their characteristics, particularly on US or MRI, have not been studied systematically.
Objectives. To provide an overview of the clinical and imaging features of the different disorders, and to correlate them with the currently used classification schemes.
Material and methods. Twenty-five patients with fibrous tumours were evaluated retrospectively. Clinical histories were studied for the histopathological diagnosis, age, signs and symptoms at presentation, mode of therapy and follow-up where available. Imaging findings were analysed for the following variables: number, location, size, margin and architecture of soft tissue and/or visceral lesions and the presence and pattern of osseous involvement. Comparison with the available literature was performed.
Results. The following tumour types were encountered: desmoid fibromatosis ($n = 9$), myofibromatosis ($n = 7$), fibromatosis colli ($n = 2$), congenital-infantile fibrosarcoma ($n = 2$), adult-type fibrosarcoma ($n = 2$), fibrous hamartoma of infancy ($n = 1$), angiofibroma ($n = 1$) and hyaline fibromatosis ($n = 1$).
Conclusions. While some tumours were non-specific in their clinical and radiological manifestation, others such as myofibromatosis, fibromatosis colli, fibrous hamartoma of infancy and angiofibroma exhibited a characteristic pattern which allowed a diagnosis to be made even without histology.

Introduction

Tumours of fibroblastic-myofibroblastic origin account for 12% of soft tissue neoplasms in childhood, being fourth in frequency after vascular, neural and rhabdomyomatous tumours [1]. Based on histopathological and clinical criteria, many types of childhood fibrous tumours are distinguished (Table 1) [1–4]. Many are diagnosed in neonates, infants and young children. Benign tumours are called fibromas or fibromatoses, while malignant tumours are known as fibrosarcomas.

Material and methods

To highlight the value of imaging in the diagnostic approach to fibrous tumours in children, we reviewed the clinical histories and imaging findings of 25 patients managed at four centres. The results were compared to previous literature reports. Histological proof was available in 24 patients. Classification was based on histopathological criteria [1–4]. One patient with fibromatosis colli was diagnosed on the basis of clinical presentation, imaging findings and follow-up.

The tumours were studied with following imaging modalities: plain X-ray ($n = 15$), US ($n = 12$), CT ($n = 10$), MRI ($n = 8$), barium follow-through or enema ($n = 3$), intravenous urography (IVU) ($n = 2$) and angiography ($n = 1$). Multiple modalities were used in most patients at diagnostic work-up (51 studies in 25 pa-

Table 1 Fibrous tumours of infancy and childhood (after Enzinger and Weiss [3]) (*A* adult, *B* birth)

Histological diagnosis	Age (years)	Location	Solitary	Multiple	Recurrence	Regression
Fibrous hamartoma	B-2	Axillary and inguinal	+	–	Rare	–
Myofibromatosis	B-A	Soft tissue, bone, viscera	+	+	Rare	+
Fibromatosis colli	B-2	Sternocleido-mastoid muscle	+	Bilateral	Rare	+
Digital fibromatosis	B-2	Fingers and toes	+	+	Common	+
Infantile (desmoid) fibromatosis	B-4	Muscles	+	–	Common	–
Calcifying aponeurotic fibroma	2-A	Hands and feet	+	–	Common	+
Hyaline fibromatosis	2-A	Dermis and subcutaneous tissues	(+)	+	–	–

tients). Imaging findings were analysed for the following variables: number, location, size, margin and architecture of soft tissue and/or visceral lesions and the presence and pattern of osseous involvement.

Results

Age at diagnosis ranged from 5 days to 17 years. The male:female ratio was 1.5:1. Eight types of fibromatoses or fibrosarcomas were seen: desmoid fibromatosis ($n = 9$), myofibromatosis ($n = 7$), fibromatosis colli ($n = 2$), congenital-infantile fibrosarcoma ($n = 2$), adult-type fibrosarcoma ($n = 2$), fibrous hamartoma of infancy ($n = 1$), angiofibroma ($n = 1$) and hyaline fibromatosis ($n = 1$).

Desmoid fibromatosis

Desmoid fibromatosis was diagnosed in nine patients (five boys, four girls). Age at diagnosis ranged from 8 days to 13 years 8 months (mean 7 years 1 month). Seven tumours were unifocal at presentation (three intra-abdominal, four extra-abdominal) and two cases had multifocal disease (both with intra-abdominal mesenteric and retroperitoneal spread and ascites). Six patients had recurrent or progressive unresectable disease; two of these patients died. One of those patients had Gardner syndrome (familial adenomatous polyposis), inherited from her father. She presented at the age of 13 years with hydronephrosis due to retroperitoneal fibromatosis around the left ureter in the absence of polyps (Fig. 1).

Imaging features: The findings were non-specific on US and CT. The solid masses appeared homogeneous or mildly heterogeneous, ill-defined or well-defined, of variable echogenicity on US, and hypodense or isodense compared to muscle on CT (Fig. 1). On MR (four cases), one lesion showed mildly low, and a second lesion moderately low signal intensity on T1-weighted (T1-W) and T2-weighted (T2-W) images (Fig. 2). Two other lesions were isointense to mildly hyperintense on both sequences. A single lesion in the posterior right thigh, abutting the distal femoral diaphysis, led to a shallow indentation and irregularity of the cortex. The other tumours did not involve bone. The diameter of the lesions was variable, ranging from a few centimetres to more than 10 cm.

Two boys presented with ascites and intestinal obstruction due to multicentric mesenteric and retroperitoneal fibromatosis. The imaging findings consisted of ascites, bowel wall thickening and evidence of small-bowel and colonic obstruction (Fig. 3).

Myofibromatosis

Myofibromatosis was seen in seven patients (five girls, two boys). Age at diagnosis ranged from 5 days to 4 months (mean 6 weeks). Two patients had unifocal disease and five patients had multifocal involvement of bones, muscles, subcutaneous tissues and viscera (adrenal). Small-bowel leiomyomatosis was also present in one case. Complete regression was documented in three cases (observation period 4 months to 9 years 7 months). In two cases, a combination of partial regression and progression (in size and number) was noted within 2–7 months. One patient showed progressive disease during 3 months follow-up. One unifocal tumour was excised.

Imaging features: Soft-tissue lesions were rounded and well defined on US and MRI (Fig. 4a–c). The tu-

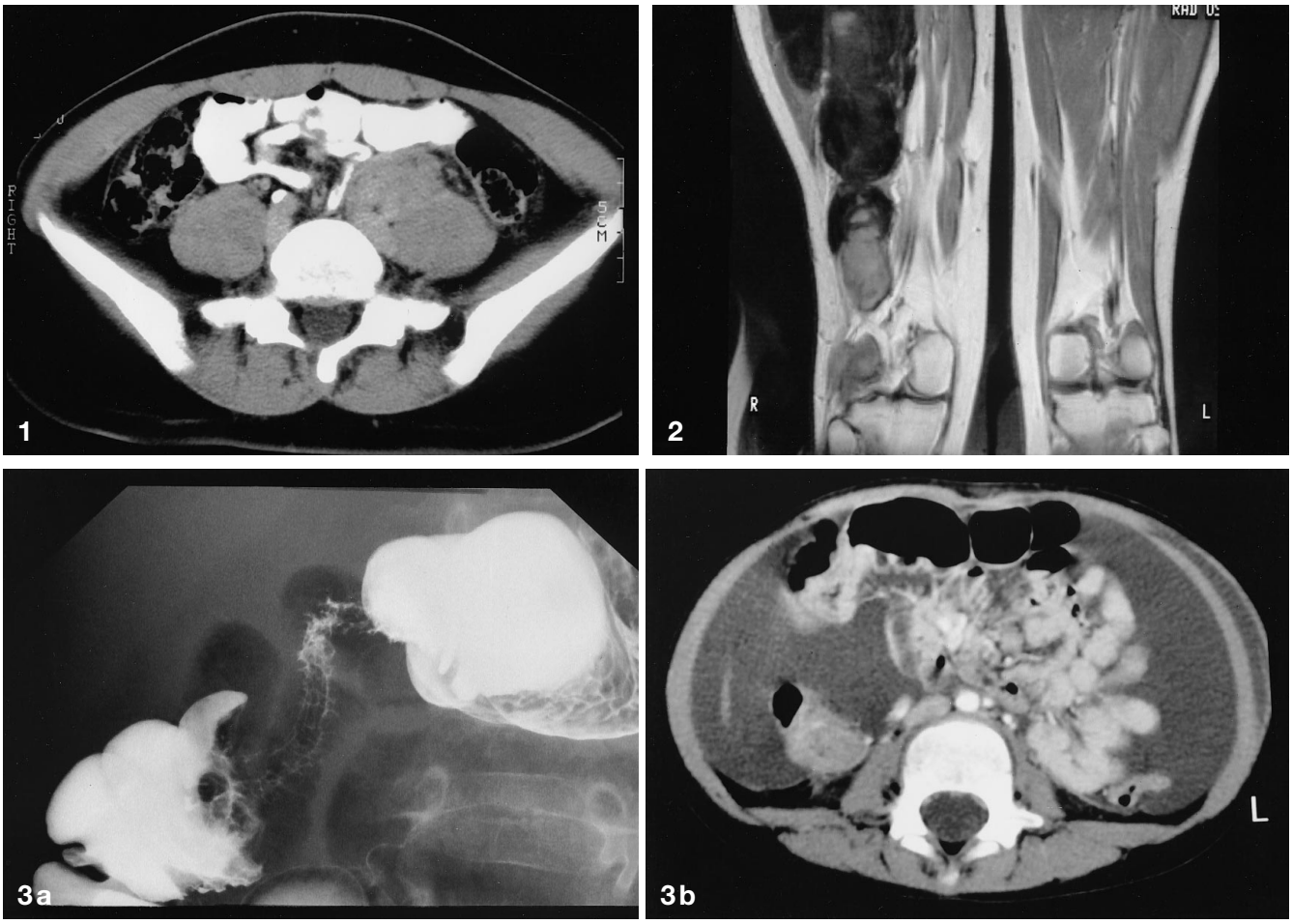

Fig. 1 A 13-year-old girl with Gardner syndrome and retroperitoneal desmoid fibromatosis. CT demonstrates an enhancing and relatively homogeneous mass anterior and medial to left psoas muscle with displacement of bowel loops. The left ureter was obstructed and the descending and sigmoid colon were infiltrated by the tumour

Fig. 2 A girl with recurrent desmoid fibromatosis of her right thigh. Follow-up MR (SE 1500/20) at age 18 years shows a hypointense tumour of biceps femoris muscle

Fig. 3 a,b A 5-year-old boy with multicentric mesenteric and retroperitoneal fibromatosis. **a** Contrast enema showing circumscribed narrowing of the hepatic flexure. **b** Contrast-enhanced CT demonstrates massive ascites and bowel wall thickening of the ascending and proximal transverse colon

mours measured from less than 1 cm to a few centimetres in diameter. On MR, the lesions were isointense to muscle on T1-W images, hyperintense to muscle on T2-W images and strongly enhancing with contrast medium. Some of the lesions showed a characteristic target sign appearance on US and MRI. The centre of these lesions was mildly hyperintense on T1-W images and non-enhancing. Central calcification of soft tissue and skeletal tumours was observed on XR in two cases on follow-up examination (Fig. 4 d). Osteolytic skeletal involvement was documented in the skull, spine, chest, shoulder girdle, pelvis and extremities. Within the tubular bones, lesions were located predominantly in the metaphyseal region, less often in the diaphysis. The central or eccentrically located lesions were lytic, sharply marginated and sometimes expansile, with or without cortical disruption. A sclerotic rim developed with regression (Fig. 4 d). In two cases, the spine showed multifocal involvement of vertebral bodies and posterior elements with variable vertebral collapse and vertebra plana (Fig. 4 e).

Fibromatosis colli

Fibromatosis colli was diagnosed in two boys aged 3 weeks and 2 months. Both patients presented with a swelling in the lateral aspect of the neck, and one had a torticollis. One patient had been delivered by caesarean section because of breech presentation, while vacuum extraction had been performed in the other. The

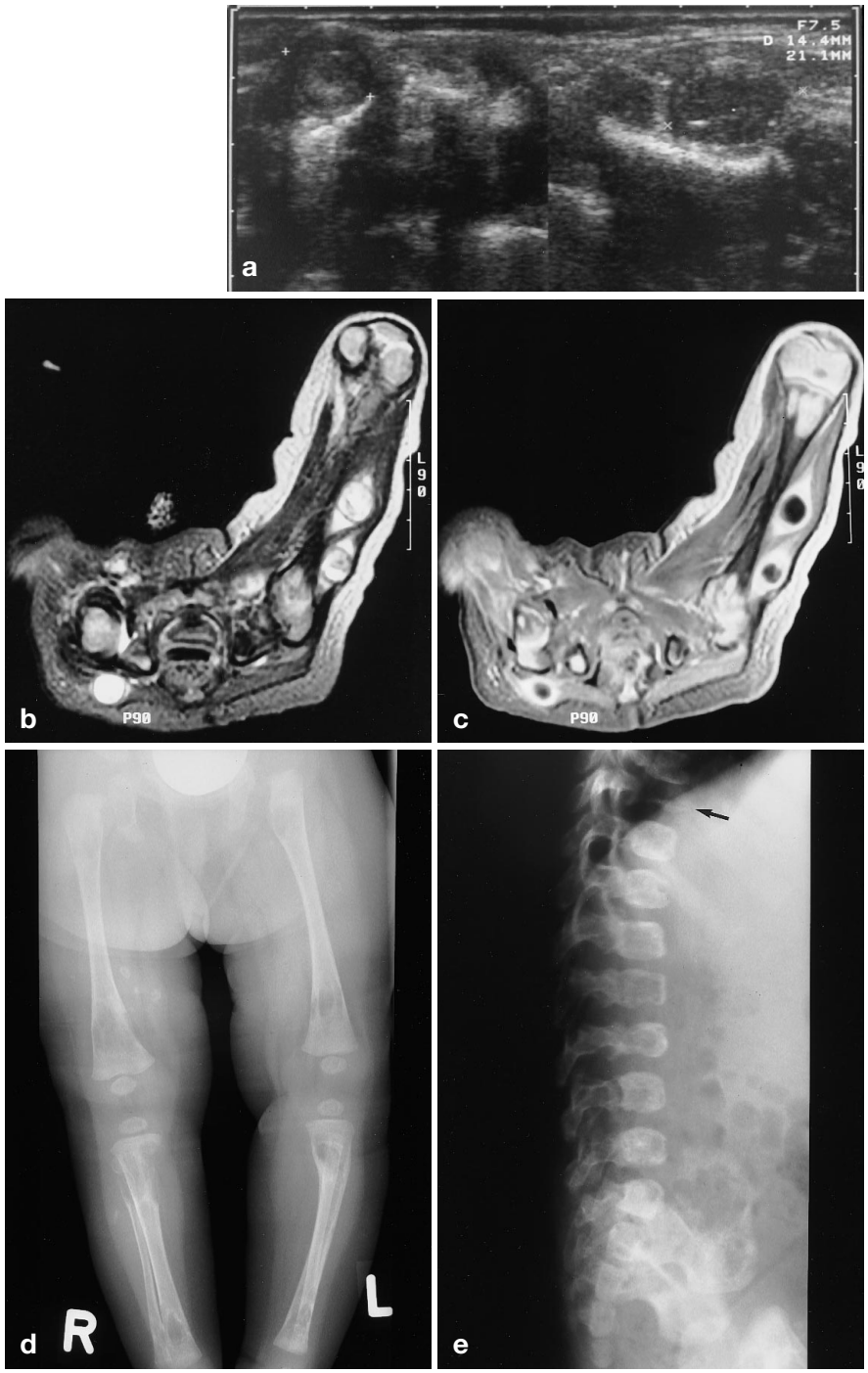

Fig. 4a–e A baby girl with myofibromatosis. **a** US shows two different soft-tissue lesions with a target sign appearance. **b** MR (FSE 5400/102 with fat saturation) of the pelvis and left thigh shows marked hyperintensity of the lesions. **c** MR (SE 540/16 with gadolinium enhancement and fat saturation) demonstrates centrally non-enhancing and peripherally enhancing intramuscular lesions. Note the enhancing metaphyseal lesions in the left femur. **d** XR of legs shows characteristic metaphyseal (and diaphyseal in the left fibula) osteolytic lesions with sclerotic rims. Note calcification of the soft tissue tumours. **e** XR of the lumbar spine shows vertebra plana of T10 (*arrow*). Multiple lumbar vertebral bodies are partially destroyed

tumour was resected in one case because of suspected malignancy. The other patient was followed clinically.

Imaging features: The tumours appeared as central (spindle-shaped) or exophytic enlargement of the sternocleidomastoid muscle on US (Fig. 5 a). The lesions were mildly heterogeneous and hypoechoic or isoechoic relative to muscle. On CT (one case), the tumour was isodense to mildly hypodense relative to muscle on unenhanced and enhanced scans (Fig. 5 b). The lesions measured several centimetres in diameter.

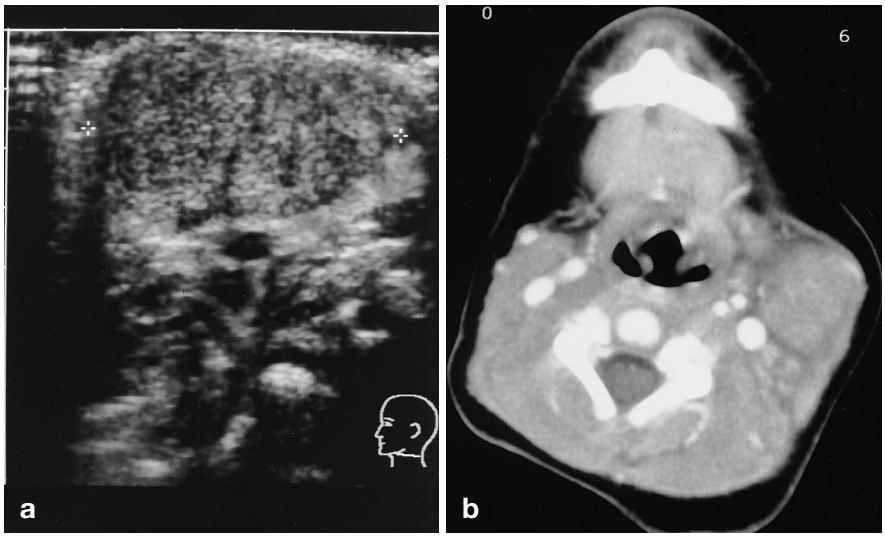

Fig. 5a,b Fibromatosis colli in a 3-week-old boy. **a** US (transverse section) of the left side of the neck showing a mildly heterogeneous mass arising from the sternocleidomastoid muscle. **b** CT shows an isodense mass during contrast injection. Note the displaced and elongated sternocleidomastoid muscle

Congenital-infantile fibrosarcoma

Congenital-infantile fibrosarcoma was seen in two infants (one boy, one girl) 3 and 6 months of age. They presented with a large, solitary mass in the axilla and calf, respectively. Both masses decreased in size, one during chemotherapy and one following cessation of chemotherapy (Fig. 6a).

Imaging features: The tumours measured 5 cm and 8 cm, respectively. Osseous erosion of the adjacent bones was noted in both (Fig. 6a). Relative to muscle, the tumours were isoechoic on US and mildly hypodense on unenhanced CT. On MR, they appeared isointense on T1-W sequences and hyperintense on T2-W images. The enhancement pattern was heterogeneous, with areas of low signal intensity, both centrally and in the periphery of the tumours (Fig. 6b).

Adult-type fibrosarcoma

Adult-type fibrosarcoma was diagnosed in two boys 15 and 17 years of age. The tumours presented with pain due to an intraosseous tumour of the calcaneus and as a soft tissue mass in the forearm.

Imaging features: CT in one case showed a lytic, sharply marginated soft-tissue-density lesion of the calcaneus with periosteal elevation and disruption. Arteriography of the forearm in the other case revealed a richly vascularised tumour containing tortuous and irregular vessels without osseous involvement (Fig. 7). Both lesions measured 8 cm in diameter.

Fibrous hamartoma of infancy

A painless lump in the upper arm of a 9-month-old boy was diagnosed as fibrous hamartoma of infancy.

Imaging features: XR, US and MR showed a solitary subcutaneous lesion in the medial aspect of the right upper arm measuring 4 cm in its longest diameter. On US, the lesion was hyperechoic, with poorly defined margins and areas of acoustic shadowing. Calcifications were absent on XR. MR showed a characteristic mixture of fat and intermediate intensity tissues, which were isointense on T1-W and mildly hyperintense to muscle on T2-W images (Fig. 8).

Angiofibroma

This tumour was discovered in a 15-year-old boy because of fullness in his left cheek without nasal obstruction or epistaxis.

Imaging features: Both CT and MR showed a homogeneously enhancing mass in the pterygopalatine fossa with extension into the nasal cavity, the sphenoid sinus, and the base of the skull. The maxillary sinus was displaced anteriorly (Fig. 9). The mass was hyperintense on T2-W images with multiple serpiginous flow voids.

Hyaline fibromatosis

Hyaline fibromatosis (infantile systemic hyalinosis) was suspected clinically and confirmed on a skin biopsy in a 14-month-old boy who presented with joint contractures, stiff and locally discoloured skin, chronic pain and failure to thrive from early infancy. Later, subcutaneous swellings developed at both wrists and at other lo-

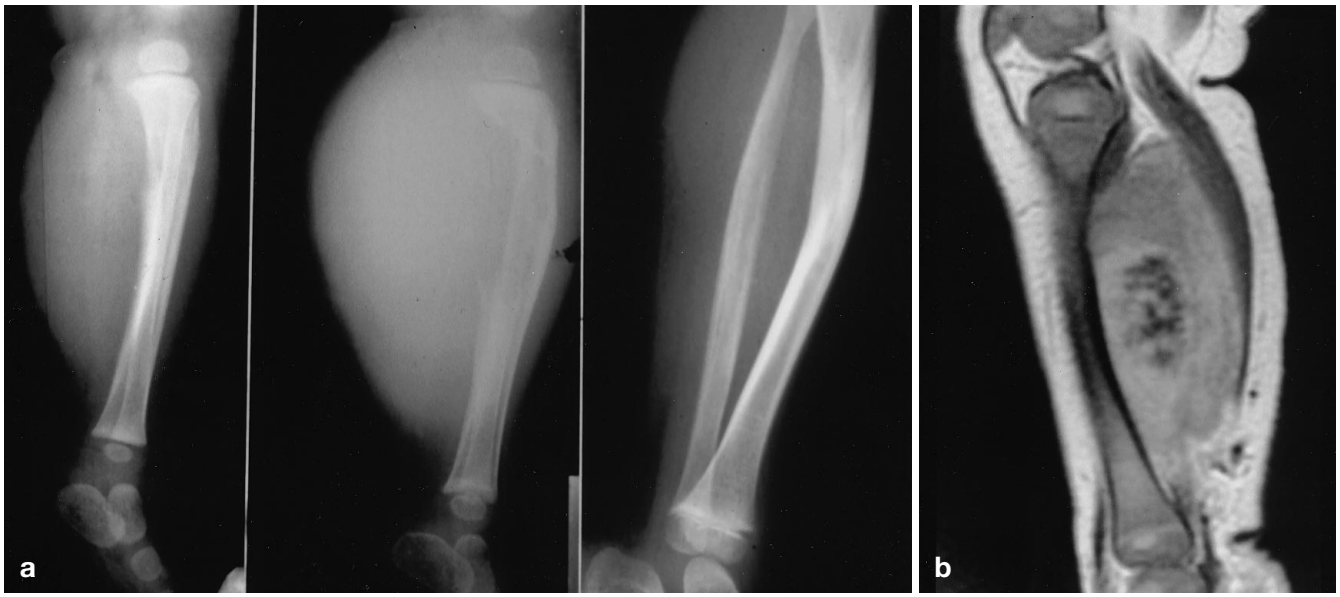

Fig. 6 a,b Congenital-infantile fibrosarcoma in a girl. **a** Evolution of the soft-tissue calf mass at 6, 13 and 30 months. Note the osseous destruction and remodelling, and regression of the tumour following cessation of chemotherapy at 10 months of age. **b** Gadolinium-enhanced MR (400/25) showing peripheral enhancement and central non-enhancement of the large mass which abuts the tibia and displaces the muscles of the calf posteriorly

cations, including as small papules in the perianal region and gingival hyperplasia.

Imaging features: Marked osteopenia, particularly at the metaphyses of long bones, flexion contractures of joints and subcutaneous soft-tissue lesions at both wrists were noted in our patient (Fig. 10 a). The carpal bone age was accelerated. A depression was noted on the medial side of the proximal tibial metaphysis (Fig. 10 b).

Discussion

A tumour predominantly composed of myofibroblasts or fibroblasts is called a fibroma or fibromatosis if it appears benign on histological examination, or a fibrosarcoma if the pathological criteria for malignancy are fulfilled. Differentiation has to be made from other similar appearing myofibroblastic lesions that are considered pseudoneoplastic, such as nodular fasciitis or inflammatory pseudotumour [1]. The term fibromatosis is used for fibroblastic-myofibroblastic proliferations with the following characteristics [1, 3, 4]:

1. A tendency to invade surrounding tissues
2. A tendency to recur after incomplete excision
3. Absence of metastases

In addition, some fibromatoses may show the following features:

4. Spontaneous regression
5. Presentation with multifocal tumours

In general there is a high incidence of fibromatoses in neonates, infants and young children.

The most widely accepted classification scheme for fibrous tumours was proposed by Enzinger and Weiss [3], who divided paediatric fibrous tumours into two categories:

1. Tumours 'corresponding in clinical setting, microscopic picture, and behaviour to similar lesions occurring in adults', including desmoid fibromatosis
2. Tumours 'that are peculiar to infancy and childhood and generally have no clinical or morphological counterpart in adult life' (Table 1)

Other authors expanded the spectrum of fibroblastic-myofibroblastic tumours in children and adolescents using other clinical or pathological criteria, or weighting the criteria differently. They added angiofibroma, congenital-infantile and adult-type fibrosarcoma, and fibroma of tendon and nerve sheath [1, 2].

Desmoid fibromatosis

Desmoid fibromatosis is a term used synonymously with aggressive, infantile or musculoaponeurotic fibromatosis [1, 3]. Paediatric cases are diagnosed in the first and second decades of life, more frequently in boys than girls [1, 4]. Familial cases occur, particularly as part of

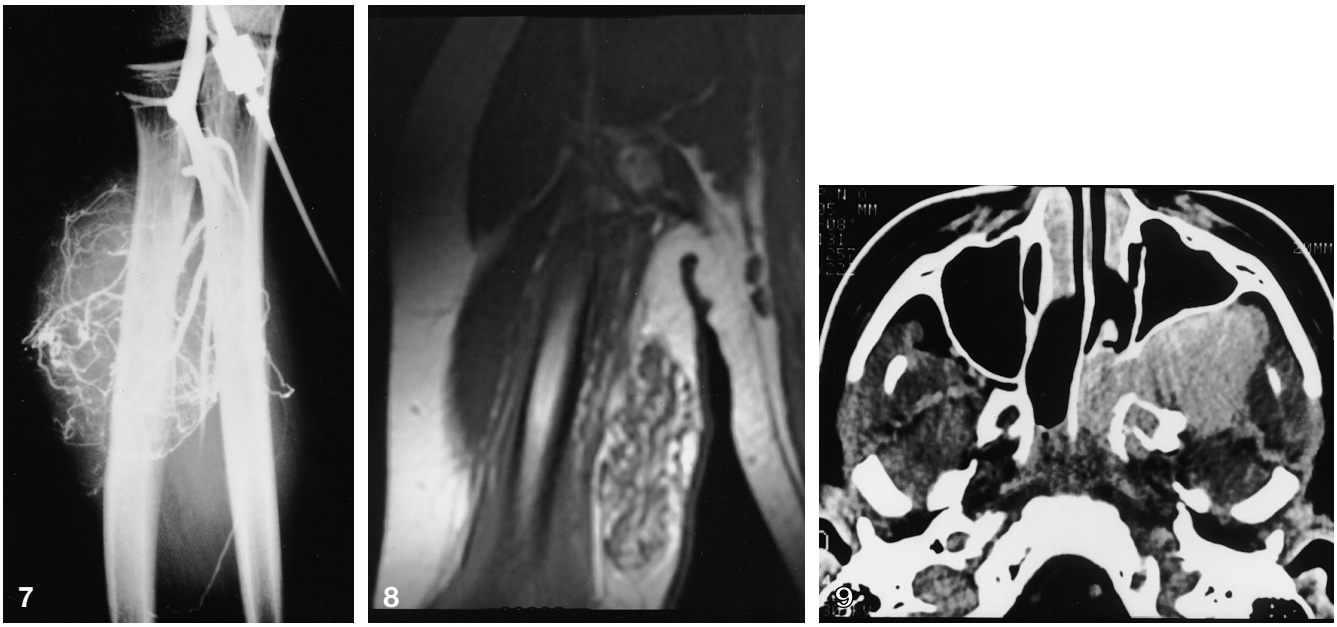

Fig. 7 Adult-type fibrosarcoma in a 17-year-old boy. Arteriography of the forearm (arterial phase) reveals a richly vascularised tumour containing tortuous and irregular vessels, but no osseous involvement. The brachial artery and the bifurcation are displaced by the mass. The tumour stained uniformly. No arterio-venous shunts were seen

Fig. 8 Fibrous hamartoma of infancy in a 9-month-old boy. MR (500/15) shows a subcutaneous, oval-shaped heterogeneous (fibrous and fat tissue) mass on the medial aspect of the proximal right arm. Note the regular 'organoid' structure

Fig. 9 Angiofibroma in a 15-year-old boy. CT shows a homogeneously enhancing mass arising from and expanding the left pterygopalatine fossa. There is invasion of the nasal cavity. The posterior wall of the maxillary sinus is displaced anteriorly

Gardner syndrome (autosomal dominant inheritance), and presentation may be with multifocal lesions. Development of the fibromatosis may be related to previous surgery or radiation. The biological behaviour is variable, with one or multiple recurrences being documented in about 60% of patients. Death due to uncontrolled local growth can occur [1, 4].

Radiographs may demonstrate osseous involvement with erosion, deformation and bowing of bone adjacent to the tumour [3]. CT usually shows a soft-tissue-density lesion (Fig. 1), but high-density tumours have been reported in adults [5]. MR shows intermediate or low T1 signal intensity and high or low intensity on T2-W images (Fig. 2). Hypointensity on T2-W images has been shown to be due to a greater collagenous component and reduced cellularity compared with lesions showing high signal intensity [6]. A nodular pattern is more frequently present in adults, while paediatric patients more often show an infiltrative pattern, which is associated with a higher risk of recurrence [7]. Bone scintigraphy has a certain value in detecting multifocal lesions, but sensitivity and specificity, particularly concerning soft-tissue lesions, appear to be poor [3]. Bowel wall thickening may be a sign of mesenteric fibromatosis. Ascites can accompany mesenteric involvement due to lymphatic and venous obstruction, or hypoproteinaemia secondary to protein-losing enteropathy (Fig. 3) [8].

Myofibromatosis

Myofibromatosis (congenital generalised fibromatosis) was the second most frequent diagnosis in our series. A slight male predominance has been reported [9]. Myofibromatosis presents most often in infancy, but can occur even in adults. The solitary or multiple lesions involve skin, subcutaneous tissues, muscles, bones and viscera (particularly lung, heart, gastrointestinal tract and dura) [1, 9]. Multicentric visceral involvement, especially of the lung, is considered a poor prognostic indicator associated with early death [9]. The tumours measure up to 8 cm in diameter and occasionally contain a central focus of haemorrhage, cystic degeneration, necrosis, calcification, ossification and fat [1], which explains the target sign appearance on US and MR. Evolution consists of an increase in size and number of lesions up to 1 year, followed by a regressive phase over the ensuing years. In survivors, healing without residual skeletal deformity can be expected. The recurrence rate of excised lesions is low [9, 10]. XR findings of multicentric myofibromatosis are characteristic. The skeletal lesions appear lytic and sharply defined. Sclerotic margins appear later, probably indicating the reaction to shrinking le-

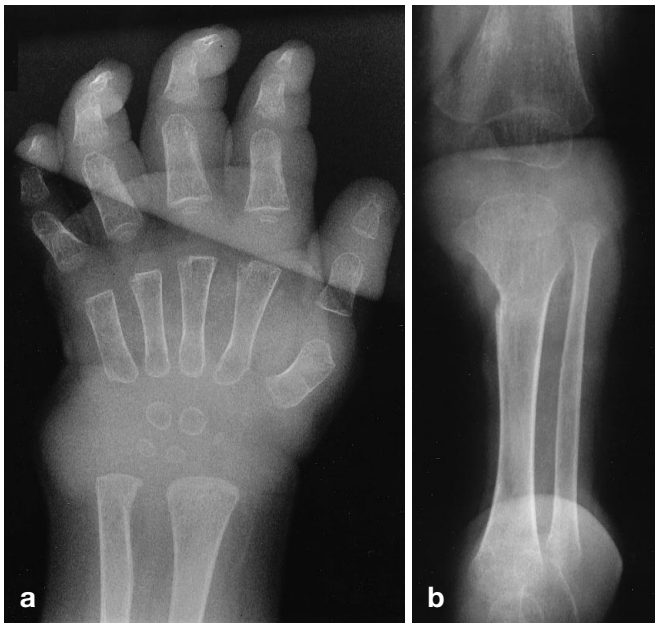

Fig. 10 a,b Hyaline fibromatosis in a 14-month-old boy. **a** Soft tissue tumours at the wrist and distal radio-ulnar joint and premature ossification of carpal bones. There are flexion contractures of the fingers. **b** Marked osteopenia is present, particularly in the metaphyses. The knee is flexed and there is a depression at the proximal tibial metaphysis medially, resembling the features of focal fibrocartilaginous dysplasia

sions during the healing phase (Fig. 4) [9, 10]. US, CT and, particularly, MR help in defining the distribution of soft tissue and visceral lesions [11]. Vertebra plana can be an incidental finding (Fig. 4) [12].

Fibromatosis colli

Fibromatosis colli presents as a mass of the lateral neck in neonates and young infants. The diameter of the lesion measures up to 3 cm. Fibromatosis colli is often associated with birth trauma and torticollis. There is a strong male predominance. Spontaneous regression is the rule. Recurrences are unusual [1, 3, 4]. The diagnosis is straightforward in the appropriate age group, and can usually be made clinically. Imaging, particularly US, can help in demonstrating the origin of the mass in the sternocleidomastoid muscle and in excluding mimicking conditions such as cervical lymphadenopathy. Fibromatosis colli appears as a focal or diffuse, relatively homogeneous enlargement of the sternocleidomastoid muscle (Fig. 5). It can be shown to move with the muscle. Other soft tissue masses of the neck (particularly rhabdomyosarcoma or neuroblastoma) should be suspected if the clinical setting is not appropriate for the diagnosis, or if imaging studies show a heterogeneous structure of the lesion, an exophytic growth, an irregular margin or regional lymphadenopathy. In such circumstances, biopsy of the lesion or resection might be indicated [13]. A lytic lesion of the clavicle at the insertion site of the fibrosed sternocleidomastoid muscle may be demonstrated by radiographs, which, however, are rarely performed in the setting of a neck mass [14].

Congenital-infantile fibrosarcoma

Congenital-infantile fibrosarcoma (congenital-infantile fibrosarcoma-like fibromatosis) is considered to be a separate entity from adult-type fibrosarcoma due to its more favourable biological behaviour, despite close histological resemblance. The tumour presents predominantly in neonates and infants and occurs more frequently in boys than girls. The solitary mass is usually large, with a diameter up to 20 cm. Focal haemorrhage and necrosis may be present. The tumour can recur but rarely metastasises. Death can result from unresectable disease [1, 3]. One tumour in our series, after having continued to grow during chemotherapy, regressed following cessation of the treatment. MR showed a non-specific, large heterogeneous mass in our two cases. Partial destruction, bowing and remodelling of adjacent bones were present in both cases (Fig. 6).

Adult-type fibrosarcoma

Adult-type fibrosarcoma is a malignant potentially lethal tumour capable of recurrence and metastases. Histological differentiation from other benign or malignant spindle cell tumours, particularly the congenital-infantile fibrosarcoma is often difficult. In children the tumour occurs in the second decade and usually originates in the soft tissues of the extremities, measuring up to 10 cm in diameter. A deep-seated tumour can cause periosteal reaction or destruction of the adjacent bone. Foci of calcification or ossification may be present. Imaging features are, otherwise, non-specific except for angiography which, according to literature reports, allows the distinction between benign and malignant fibrous tumours (Fig. 7) [1, 3, 15].

Fibrous hamartoma of infancy

Fibrous hamartoma of infancy (subdermal fibrous tumour of infancy) is a tumour of neonates, infants, and young children occurring predominantly in males. This painless lesion is located in the subcutaneous tissue, most often in the shoulder girdle, less frequently in other regions, but never in the hands and feet. Adjacent structures can be infiltrated. Lesions usually measure

between 3 cm and 5 cm in diameter. Multicentric growth is rare. No metastases are recorded. Histology consists of an 'organoid' mixture of three components: fibrous tissue, adipose tissue and nests of immature mesenchyme [1, 3, 4, 16]. Local resection of the lesion is usually curative with a low risk of recurrence. MR findings are characteristic – strands of intermediate signal intensity (fibrous) tissue interspersed with fat in an organised fashion (Fig. 8) [17].

Angiofibroma

Angiofibroma is a benign, fibrovascular neoplasm that occurs almost exclusively in male adolescents. The tumour typically originates in the nasopharynx near the pterygopalatine fossa. Epistaxis or nasal obstruction are the leading symptoms. Locally aggressive growth may lead to spread into the adjacent paranasal sinuses, nasal cavity or infratemporal fossa. Further extension through the inferior orbital fissure into the orbit and intracranially through the foramen rotundum, pterygoid canal and superior orbital fissure is possible. Recurrence rates of up to 60 % have been reported [1, 3, 18]. Imaging findings are characteristic, showing a well-defined homogeneous and strongly enhancing mass in the area of the pterygopalatine fossa (Fig. 9). The lesion is hyperintense on T2-W images. Flow-void channels within the tumour represent vascular structures [18]. Angiography serves for confirmation of the diagnosis, planning of treatment and embolisation as a definitive procedure or prior to surgery.

Hyaline fibromatosis

Hyaline fibromatosis and infantile systemic hyalinosis most likely represent different expressions of the same disorder, infantile systemic hyalinosis being the more severe with early onset and worse prognosis, and juvenile hyaline fibromatosis being a milder form with later onset. Autosomal recessive inheritance is likely [19]. Patients with the infantile form show stiff skin, chronic pain, recurrent infections, failure to thrive and bloody diarrhoea, and die early. Children affected with juvenile hyaline fibromatosis generally survive into adulthood and suffer from subcutaneous nodules and tumour masses up to 5 cm in diameter in the head, back and extremities. Overlapping features are flexion contractures of large joints, multiple cutaneous papules and gingival hyperplasia [3, 19]. On histology, an amorphous eosinophilic substance is found in the dermis, in gut mucosa and in the tumours. Musculoskeletal imaging shows osteopenia, particularly in the metaphyses, osteolytic lesions of skull, long bones and phalanges, and a depression at the proximal tibial metaphysis medially, which resembles the changes associated with focal fibrocartilaginous dysplasia [20]. The soft tissue tumours are non-specific on XR or US (Fig. 10).

Conclusion

Imaging of fibrous tumours in children serves multiple purposes:

1. Confirmation of a clinically suspected mass
2. Characterisation of the mass with regard to location, extension, structure and margins
3. Determination of either unifocal or multifocal disease, or metastases
4. Observation of additional features as part of a systemic disorder (e. g. Gardner syndrome, hyaline fibromatosis)
5. Formulation of a tentative diagnosis
6. Assistance for biopsy or excision
7. Follow-up for documentation of regression, progression, recurrence or secondary spread

Each of these factors is important in the management of children affected with such a disorder. However, by recognising their characteristic features, it may be possible to avoid biopsy or excision in certain cases. Fibromatosis colli and multicentric infantile myofibromatosis, both disorders with a tendency for spontaneous regression, are sufficiently distinctive in their clinical and radiological appearances to allow a confident diagnosis. Other disorders like fibrous hamartoma of infancy and angiofibroma show equally distinctive features, but may have to be excised because of progressive growth or complications. The imaging features of desmoid fibromatosis and the fibrosarcomas are non-specific, except for hypointensity on T1-W and T2-W images in desmoid fibromatosis. Hyaline fibromatosis may be suggested primarily on the basis of the clinical presentation and the radiographic features, but biopsy may have to be performed to clarify the prognosis.

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