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CASE REPORT

Fibrous hamartoma of infancy mimicking teratoma in the parapharyngeal space on multidetector row CT

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Abstract To the best of our knowledge, fibrous hamartoma of infancy (FHI) mimicking teratoma presenting in the parapharyngeal space has not been reported to date. We present one case of this tumour and describe its characteristic findings on multidetector-row CT (MDCT). This case is of interest in that the parapharyngeal space was involved and the mass mimicked teratoma on MDCT. We present the MDCT findings and briefly review the relevant literature.

Keywords Fibrous hamartoma · Teratoma · MDCT · Child

Introduction

Fibrous hamartoma of infancy (FHI) is an uncommon benign tumour usually occurring in the first 2 years of life. The lesion frequently arises in the upper part of the body including axilla, upper arm and neck. Isolated cases involving the finger, foot, perianal region, scrotum and oropharynx have also been reported [1–6]. However, FHI presenting in the parapharyngeal space has not been reported to date. We report a case of FHI occurring in the parapharyngeal space and describe the characteristic manifestations on multidetector-row CT (MDCT).

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Case report

A 10-year-old boy presented with a history of snoring since birth. The symptom had become gradually worse since the age of 6 years and was associated with breath-holding when sleeping. Physical examination revealed a swollen left tonsil. Routine laboratory findings were within normal range. MDCT scan (parameters: 120 kVp, 115 mAs; CT dose index 7.63 mGy) showed a well-defined mass in the left parapharyngeal space, measuring approximately 5.0×4.0×4.0 cm in size. The lesion was of heterogeneous CT density with the centre of the mass of soft-tissue density with dot-like calcification encapsulated by fatty tissue (Fig. 1). The vessels and muscles in the vicinity were compressed, but no bony or visceral involvement was revealed. On the basis of these findings, teratoma was suspected before operation. Local excision was performed under anaesthesia. A well-demarcated tumour in the left parapharyngeal space was identified and removed. Histopathological examination confirmed the mass was composed of bundles of fibrous tissue, primitive mesenchymal cells and mature adipose tissue islands (Fig. 2). Based on these findings, a final diagnosis of FHI was made.

Discussion

FHI is a relatively rare, benign soft-tissue tumour first described by Reye [1] in 1956 as a subdermal fibromatous tumour of infancy and named FHI by Enzinger [2] in 1965. FHI are more frequently found in boys than girls and in those younger than 2 years of age. However, Efem et al. [3] reported a 10-year-old girl who presented with a solitary lesion in left supraclavicular fossa. We report a case occurring in a 10-year-old boy.

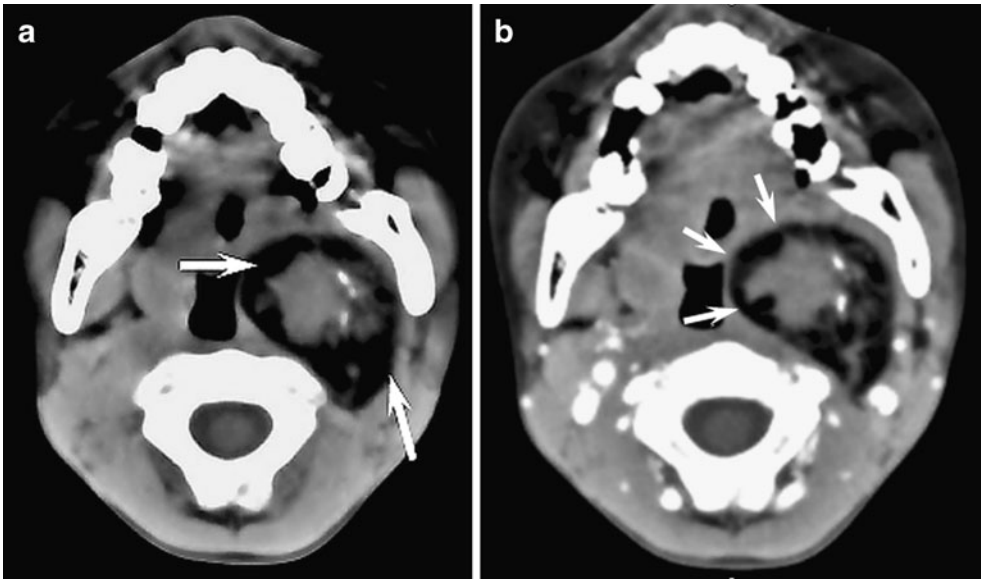

Fig. 1 FHI in the left parapharyngeal space in a 10-year-old boy. **a** Axial unenhanced CT shows a large mass in the left parapharyngeal space, measuring 5.0×4.0×4.0 cm in size, with dot-like calcification in the soft-tissue central area and peripheral fatty tissue (*white arrows*). **b** Contrast-enhanced CT shows a well-defined mass (*white arrows*) with heterogeneous density and without contrast medium enhancement

Most lesions present as a firm, nontender subcutaneous mass, usually slow-growing, although tumours growing rapidly from birth to the age of about 5 years have been reported. The common sites of involvement are the axilla, inguinal region, upper arm, chest wall and neck. FHI arising in the parapharyngeal space is extremely uncommon. To the best of our knowledge, this is the first reported case that includes the accompanying CT features.

Previous studies described the characteristic histopathological features as follows: (1) bundles of fibrous tissues in varying proportions; (2) foci of collagen-poor, moderately or densely cellular myxoid mesenchymal tissue, and (3) islands of mature adipocytes [2, 3]. The varying proportions of the three types of components represent the different imaging features on US, CT and MRI. Ashwood et al. [4] and Loyer et al. [5] described the MRI features of the lesions and correlated these with the pathological findings. MRI is usually recommended for assessing paediatric soft-tissue tumours as excellent tissue characterization accurately reflecting the histological components of FHI is achieved with the absence of ionizing radiation. The fat components of FHI are hyperintense on T1-weighted images and of reduced signal intensity on T1-W images with fat suppression. The fibrous tissues are usually hypointense on T1- and T2-W images.

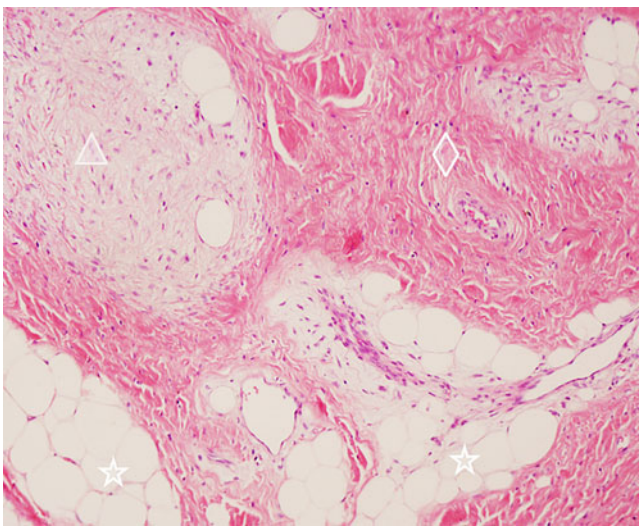

Fig. 2 Photomicrography of histology specimen shows the tumour consists of bundles of fibrous tissue (△), primitive mesenchymal cells (◇) and mature adipose tissue islands (☆) (haematoxylin & eosin, × 100)

US is a nonspecific modality for FHI. Usually, US reveals a mixed echogenicity lesion that may be a lipoma, haemangioma or teratoma. Stock et al. [6] described the US findings of a FHI in the scrotum with homogenous echo pattern similar to the testes. However, the CT features of this tumour have rarely been reported in the literature. For the first time, we report an uncommon tumour locating in the parapharyngeal space mimicking a teratoma on CT images. In the present case, the characteristic CT manifestations of this lesion included the fat density and dot-like calcification. With MDCT, two-dimensional multiplanar and three-dimensional reconstruction images provide an important method to evaluate the mass and the involvement of the vital adjacent neurovascular structures, and offer the potential to improve diagnostic confidence and accuracy so as to avoid unnecessary biopsy.

Although the histopathological features of FHI are well-described and characterized, the differential diagnosis of this entity based on imaging has not been clearly described due to its rarity. The differential diagnosis lipoma, liposarcoma, fibrolipomatous hamartoma. In common, a fatty component is suggestive of a benign lesion in infants. Ashwood et al. [4] reported that FHI was

vascular with more large vessels coursing around and through it than is normally seen with simple lipomas. In addition, compared with lipomas, the signal of FHI was only partially suppressed by a fat suppression inversion sequence. Lipoblastoma is a relatively rare benign adipose tumour that occurs in infancy and early childhood and arises from embryonic white fat, most commonly involving the extremities. Fibrolipomatous hamartoma is a rare lesion frequently affecting the median nerve. When a well-demarcated tumour with no sign of involvement of surrounding structures is identified, a benign mass should be considered in order to differentiate the lesion from a liposarcoma. As malignant adipose tumours, liposarcomas are reported in young adults and teenagers. However, liposarcomas account for <5 % of all soft-tissue sarcomas in children and are not observed in children younger than 10 years of age. Furthermore, CT is sensitive for detecting calcific foci. As soft-tissue calcification is rarely detected in FHI, teratoma was initially suspected in this case. Although extremely rare, FHI should be considered in the differential diagnosis of a tumour containing adipose tissue when calcification within the mass has been detected.

Preoperative assessment of FHI with advanced imaging modalities is possible; however, definitive diagnosis depends on histological examination. Although the diagnosis of the fatty tumours of infancy with biopsy is "nonspecific," fine-needle aspiration has been recommended as a safe and effective technique in the evaluation of FHI [7]. Local excision was the first treatment of choice because a benign clinical course was shown by long-term follow-up, but Enzinger et al. [2] reported a 16% local recurrence rate due to incomplete excision. CT may be helpful to establish a surgical operative plan to excise adequately the lesion with the aim of preventing local recurrence.

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