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CASE REPORT

Embryonal carcinoma in a cryptorchid testis of a 3-year old

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Abstract The undescended testis is at an increased risk of malignant transformation. Almost all such tumours occur in the second to fourth decades of life and are usually seminomas. A case of a mixed germ-cell tumour with yolk sac and embryonal carcinoma components arising in one of the hitherto uncorrected bilateral cryptorchid testes of a 3-year old child is presented.

Keywords Cryptorchidism · Embryonal carcinoma · Prepubertal

Introduction

The propensity of the undescended testis (UDT) to undergo malignant change is well established and discussed widely in literature. Malignant degeneration commonly takes place in the third and fourth decades of life and the common tumour to arise in the cryptorchid testis is usually a seminoma. We report a rare histological variant of a tumour in a 3-year-old child with bilateral intraabdominal testis.

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Case report

A 3-year-old child presented with lower abdominal pain, since a fortnight and a hypogastric mass noticed since 1 week. Born by a term normal vaginal delivery, he had an uneventful antenatal and perinatal period. He was noticed to have bilateral non-palpable undescended testes at birth and advised follow-up; however, the parents had not sought further medical attention till they reported here. Abdominal examination revealed a solitary, firm, non-tender mass occupying the hypogastrium and bilateral iliac fossae. The mass had a limited transverse mobility. The scrotal sacs were underdeveloped and empty whilst the penis was grossly normal.

The serum beta-human chorionic gonadotrophin (HCG) was 1.47 mIU/ml (normal 0–2 mIU/ml) and the serum alpha fetoprotein (AFP) was 250 ng/ml (normal < 10 ng/ml). The chest X-ray was unremarkable. Abdominal ultrasonography revealed a well defined, non-calcified, heteroechoic lesion; the contrast-enhanced CT showed a well defined, septated, cystic solid mass, with enhancing solid areas in the lower abdomen. The gonads were not visualised separately, there was no retroperitoneal lymph node enlargement and the fat planes between the mass and the surrounding structures were preserved. A right hydroureteronephrosis was also noted.

A provisional diagnosis of a germ-cell tumour (GCT) arising from one of the undescended testes was made. At laparotomy, a discrete 8 × 10-cm mass was seen to replace the left undescended testis. An atrophic right testis was located just cephalad to the inguinal ring. All other viscera were grossly normal and there was no ascites. A high left orchidectomy (Fig. 1) and right orchidopexy was performed.

Grossly, the tumour was well encapsulated. On section, it had a solid, variegated appearance with microcysts, foci of haemorrhage and necrosis. Microscopically, there were two distinct patterns of neoplastic cells. One showed solid

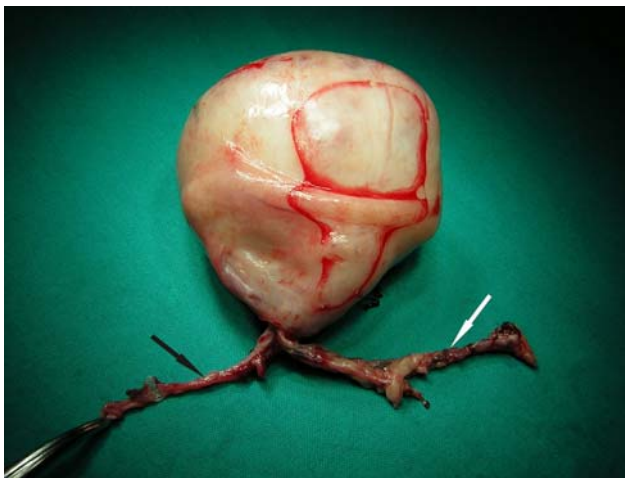

Fig. 1 Orchidectomy specimen showing well-encapsulated testicular tumour with the spermatic cord structures (*black arrow*) and the testicular vessels (*white arrow*)

sheets, nests and a glandular pattern of cells with indistinct cell margins, markedly pleomorphic nuclei and prominent nucleoli constituting the embryonal carcinoma component. The other comprised of a yolk sac tumour component with neoplastic cells having pleomorphic vesicular nuclei arranged in a microcystic and glandular–alveolar pattern. Many hyaline intra and extracytoplasmic round inclusions were seen in these areas (Fig. 2a). Extensive necrosis and haemorrhage was also noted. Immunohistochemistry for CD 30 showed focal membranous nuclear positivity in the embryonal cell component (Fig. 2b).

Postoperatively, he received one cycle of chemotherapy with bleomycin, etoposide and cisplatinum, but did not report for further treatment or follow-up, despite the repeated telephonic and postal reminders.

Discussion

Testicular tumours in prepubertal boys differ in incidence, histology and prognosis from those in postpubertal males. They occur ten times more frequently in postpubertal males than in boys 12 years and younger with a reported incidence of 0.5 per 100,000 in children. Data from the Prepubertal Testis Tumour Registry of The American Academy of Paediatrics suggest that 98% of malignant testicular GCTs in prepubertal males have pure yolk sac histology. In addition, the prepubertal malignant tumours are less likely to metastasise and have a better outcome [1, 2].

Undescended testis is a common congenital anomaly in boys; 2–5% boys born at term are born with UDT and the incidence falls to 1% by the age of a year [3]. UDT are deemed to be at an increased risk for testicular malignancy compared with those that are normally descended. The quantum of this risk has been variously quoted in different series. Swerdlow et al. [4] followed 1,075 boys with cryptorchidism managed with orchidopexy/hormones during 1951–1964 till the 1990s and estimated the relative risk (RR) of developing testicular tumours in them to be 7.5 (95% CI 3.9–12.8). A recent meta-analysis of 21 studies exploring the association of UDT with the risk of developing GCT cites an overall RR of 4.8 (95% CI 4.0–5.7) [5]. In another series of 1,335 boys, the risk of malignancy was found to be 5% when associated with an intraabdominal testis, abnormalities of the external genitalia or an abnormal karyotype against 0% in patients without these factors [6]. Although some studies indicate that there may be an increased risk of malignancy with a delay in orchidopexy, the issue remains debatable [7, 8]. However, the recommended treatment for children with cryptorchidism is unambiguous and early orchidopexy improves the potential for fertility, decreases the risks of trauma and torsion and enables an early detection of an evolving malignancy.

Although a few cases have been reported in the first decade of life, the vast majority of malignancies in UDT usually develop during puberty or later [9]. The peak age incidence of malignancy in cryptorchids is in the third or fourth decade of life irrespective of whether the testis is intraabdominal or in the groin.

The majority of these tumours are GCTs with a predominance of seminomas or mixed GCTs with elements of

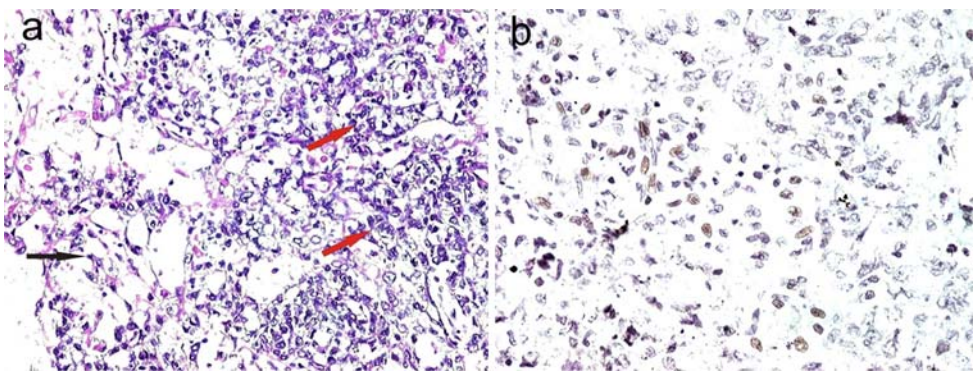

Fig. 2 **a** Microcystic pattern of arrangement of the yolk sac tumour component (*black arrow*) admixed with embryonal carcinoma showing a more solid growth pattern (*red arrow*) (H&E, ×200) **b** Focal CD 30 positivity in the cells of embryonal carcinoma (IHC ×400)

embryonal carcinomas, teratocarcinomas and choriocarcinomas [10]. Embryonal carcinoma may, in certain foci, be difficult to distinguish from yolk sac tumour when the two tumour types merge. In such cases, immunohistochemical stains aid the diagnosis. Embryonal carcinoma cells are immunoreactive for OCT 4, CD 30 and pancytokeratin whilst yolk sac tumours are immunoreactive for AFP, PLAP and low-molecular weight cytokeratin. CD 30 is highly specific for embryonal carcinoma and does not stain other GCT including seminomas and yolk sac tumours [11]. Although embryonal carcinoma cells usually show a diffuse membranous positivity for CD 30, the staining in this case was focal. A similar staining reaction has been reported by Leroy et al. [12] in 4/14 cases; also, embryonal carcinomas are generally negative for carcinoembryonic antigen, HCG and CD 117. The presence of AFP on immunohistochemical staining or in the serum, as in this case, is indicative of yolk sac differentiation in mixed tumours [11].

Mixed forms of testicular embryonal/yolk sac tumours are commoner than the pure form and are mostly encountered between 20 and 40 years and occasionally in adolescents. This variant of tumour has not been described in the prepubertal testis [13]. Generally, mixed GCTs with a predominant component of embryonal carcinoma and vascular/lymphatic invasion present in an advanced stage [13]; expectedly this tumour with a predominant yolk sac component had presented in a less advanced stage and was unusually well encapsulated.

The pathogenesis of testicular GCT is multifactorial with a variety of implicated influences. Many of these risk factors relate to prenatal influences, environmental factors or lifestyle choices [14, 15], All GCTs are thought to arise from a common precursor—the carcinoma in situ (CIS) cell. The increased incidence of CIS has been documented in the cryptorchid testis and is part of the testicular dysgenesis syndrome. These cells undergo malignant transformation in the pubertal testis under the influence of hormonal stimulation [14]. No particular predisposing factor could be identified in this case. Furthermore, the early prepubertal malignant transformation of a putative CIS cell in this testis cannot be explained with the available models of genesis of GCTs in cryptorchid testes. Probably, a hitherto unidentified intranatal/perinatal oncogenetic process could explain such an occurrence.

In conclusion, despite the rarity of such a presentation, the possibility of an early malignant transformation of a cryptorchid testis remains a distinct clinical possibility in the first few years of life.

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