Paratesticular fibrous hamartoma in an infant

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ABSTRACT Fibrous hamartoma of infancy is a rare benign tumour with local infiltration. The lesion is usually found in the upper torso and rarely occurs in the genital region. Diagnosis before surgery is rare, and complete excision is essential to prevent recurrence. We present a case in which fibrous hamartoma of infancy involving the spermatic cord was found. A pre-operative clinical diagnosis could not be made. On inguinal exploration, the tumour could not be dissected away from the testicular vessels, which necessitated an orchidectomy for complete removal. The case is presented due to its rarity and successful management.

Keywords: hamartoma, infancy, orchidectomy, tumours


INTRODUCTION

Fibrous hamartoma of infancy (FHI) is a rare soft tissue tumour that usually presents in children, mainly in boys under two years of age.1 Although the tumour has been reported to occur in different sites of the body, it is found mainly in the upper torso. An extensive review of the literature found only eight reports of FHI in the genital region and only one in the spermatic cord.2–8 We report the second case of FHI in the spermatic cord. The importance of correct diagnosis lies in determining the extent of surgery and the need for adjuvant therapy.9,10 A brief review of the diagnostic modalities and management of the condition is also presented.

CASE REPORT

A one-year-old boy born of a non-consanguineous marriage presented to the outpatient department with a slowly progressing right scrotal swelling for the past one month. The swelling was not reducible. There was no history of previous scrotal swelling, trauma, fever or loss of weight. General physical examination was normal, and there was no significant lymphadenopathy. Local examination revealed a non-tender, well-defined, hard nodular swelling measuring 3 cm x 3 cm at the root of the right scrotum (Fig. 1). The right testis was palpable separately and was pushing the raphe to the opposite side. The left testis was normal. A clinical diagnosis of right paratesticular neoplasm was made.

Haematological investigations, including haemoglobin estimation, total and differential counts and erythrocyte sedimentation rate were all normal. Mantoux test was negative. Ultrasonography revealed an ill-defined variegated mass with echogenic outlines on the posterolateral aspect of the right testis. The mass measured 2.5 cm x 2.5 cm and was hypovascular. Both testes were normal in size, shape and echotexture. An initial attempt at fine needle aspiration cytology yielded dry aspirate. A second attempt provided scanty aspirate that showed fibroblasts and inflammatory cells on microscopic examination.

Fig. 1 Pre-operative photograph shows a swelling at the base of the scrotum on the right side, resulting in shifting of the median raphe to the left side.

With the possibility of neoplastic pathology and consent for a possible orchidectomy, further exploration was conducted through an inguinal approach, which was extended to the scrotum for adequate exposure. It revealed a non-capsulated mass with well-defined margins, which densely infiltrated the Buck’s fascia over the penis, dartos and subdartos tissue, tunica albuginea and scrotal septum. The right epididymis and the adjacent part of the spermatic cord were completely engulfed by the mass, while the right testicle was distinct from the mass (Fig. 2).

As the cord structures and testicular vessels were involved in the lesion, we decided on an orchidectomy to completely excise the mass. The scrotal septum was reconstructed with local tissues and the wound was closed. Gross examination of the specimen showed a well-defined, non-capsulated, irregular, fleshy soft tissue mass (Fig. 3), which showed areas of pale grey fibrofatty tissue on the cut section. Histopathological examination demonstrated mature adipose tissue, spindle-shaped fibroblastic...
cells in a collagenous stroma and immature round mesenchymal cells, which were suggestive of FHI (Fig. 4). The child remained well without recurrence at one year post operation.

DISCUSSION

FHI is a benign, soft tissue growth, which was first described by Reye in 1956 as a "subdermal fibromatous tumour of infancy". Enzinger, in his review of 30 cases (including eight scrotal cases), described the entity distinctly and categorised the disease as a separate soft tissue tumour in children below two years of age. He termed the entity a "fibrous hamartoma of infancy". FHI is more common in boys and the median age of presentation is ten months, although 15%-20% of lesions are found at birth. The common sites of origin are the axilla, shoulder, upper back and arm. It is very rare for FHI to occur in the genital area. The lesion, especially in the genital region, is frequently reported to be misdiagnosed as lymphadenopathy, rhabdomyosarcoma or malignancies arising from neural tissue. A definitive diagnosis of FHI requires histopathological evaluation.

In a 22-year retrospective review of 15 cases of genital FHI from a major paediatric referral centre, Popek et al described the pathology of FHI in cases involving the scrotum, spermatic cord, inguinal region, pubic area and labium majus. It includes the only reported case of FHI of the spermatic cord. Grossly, the tumour appears as a firm mass in subcutaneous tissue. It is non-encapsulated, poorly circumscribed and merges into the surrounding tissues. Microscopically, the tumour characteristically contains three components; haphazardly arranged mature adipose tissue, disordered fascicles of myofibroblasts with abundant collagen and small, rounded primitive mesenchymal cells arranged in whorls in association with myxoid material and numerous small capillaries. The immunohistochemical analysis has been described in detail by Popek et al and others. The histogenesis of FHI is still unclear, but its morphology fits into the criteria for hamartoma. They tend to be poorly marginated, with fibrous tendrils extending into the surrounding soft tissue. Pathologically, the lesion is also frequently misinterpreted as infantile fibromatosis, infantile myofibromatosis or nodular fasciitis.

Scrotal lesions have been reported to involve the scrotal wall, tunica and dartos, but none have been observed to infiltrate into the testis. To date, only one case involving the spermatic cord with radical orchidectomy performed for complete removal of the tumour has been reported. We present the second case of FHI in the spermatic cord, which was similarly managed by inguinal exploration and orchidectomy. None of the other cases of FHI (including the one reported by Ritchie et al, which involved a tumour masquerading as rhabdomyosarcoma of the spermatic cord) required orchidectomy during excision of the mass, as complete excision was feasible without damaging the vas deferens and testicular vessels.

FHI is a benign tumour with no reported cases of metastasis, although rare cases of local recurrence have been reported following incomplete excision. Thus, optimal management requires complete excision. As there is no need for wide excision, adjuvant chemotherapy or radiotherapy, accurate diagnosis of FHI is essential so as to avoid overtreatment.
diagnosis by imaging has been reported. Ultrasonographic evaluation of scrotal FHI has revealed a tumour with a homogenous echotexture that is similar to the testis, and is thus not helpful in diagnosis. Magnetic resonance imaging has proven to be more promising, as it is able to differentiate between fibrous tissue and fat intensity. Loyer et al described an FHI consisting of fibrous tissue as relatively consistent with low-signal intensity, whereas fat has a high signal intensity on T1-weighted sequences, which decreases slightly on T2-weighted sequences.

We report this case due to its rarity. It is recommended that on encountering a paratesticular mass, the diagnosis of a benign pathology should be considered, and testis-sparing surgery performed. However, in difficult cases such as ours, when the tumour infiltrates the vas deferens and testicular vessels, it may be necessary to sacrifice the testis in order to ensure complete removal of the tumour and to prevent recurrence. This is the only case of FHI with epididymal involvement. The need for pre-operative evaluation with radiological investigations of paratesticular tumour in patients under two years of age is highlighted. In order to reach a definite diagnosis, inguinal exploration with histological evaluation is required, and we emphasise the necessity of a complete excision (including the fibrous tendrils extending into the surrounding soft tissue). Knowledge of the disease entity and of its differentiation from malignant tumours that require adjuvant treatment is of utmost clinical importance.

REFERENCES