Congenital Infantile Fibrosarcoma of Scalp. Is Adjuvant Therapy Essential ?

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Received: 14 February 2014 / Accepted: 27 August 2014 / Published online: 4 September 2014
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Abstract Congenital infantile fibrosarcoma (IFS) is a rare tumour. Involvement of scalp is extremely rare and only a few case reports have been published till now. The exact management of these tumours is controversial. Their counterparts which involve the limbs more commonly can be radically excised, however this option is not possible in scalp. Adjuvant radiotherapy and chemotherapy has been advocated in such cases. Local recurrences and distant metastasis have been reported inspite of such therapy. We report a rare case of scalp IFS which was present since birth and was managed by surgical excision without radio and chemotherapy. Patient is well after 10 years of follow up.

Keywords Infantile fibrosarcoma · Fibrosarcoma · Adjuvant therapy

Introduction

Congenital infantile fibrosarcomas (IFS) are very rare tumours that commonly involve the distal extremities and trunk [1]. IFS of scalp are very rare tumours with only a few cases reported in literature [2–7]. The primary treatment of congenital fibrosarcomas is wide excision with adjuvant radiotherapy and chemotherapy. Even after treatment the prognosis is not certain and distant metastasis is reported. We report rapidly growing IFS of scalp presenting at birth as an ulcerated lump over vertex which was treated with wide excision without adjuvant therapy. The patient is disease free 10 years after surgery.

Case Report

A 21 days old newborn was brought with an ulcerated mass over the vertex. The child was born normally. There was a small pea size swelling over vertex at birth. It rapidly increased in size over next 3 weeks and got ulcerated. There was frequent bleeding from ulcerated surface. On examination, the child had a large ulcerated, reddish mass over the vertex measuring 5×3 cm. It was extending across the midline and was partially covered with skin (Fig. 1a). There were no symptoms or signs suggestive of neurological dysfunction.

Magnetic resonance imaging (MRI) revealed heterogeneously enhancing extra-cranial lesion with erosion of the underlying bone but without any intracranial extension (Fig. 2a, b). The patient underwent surgery through an incision that included the lesion and a margin of healthy skin circumferentially around the lesion. The lesion was dissected off the dura that was completely intact and showed no signs of invasion. The tumor was thus excised completely along with a margin of healthy skin (Fig. 1b). The excision of the tumor left a large scalp defect that was reconstructed with rotation flap with good cosmetic results. Post op period was uneventful. The margins of excised skin were free of tumour. In view of tumour free margin with total tumor excision, patient was not given adjuvant radiotherapy or chemotherapy. The patient is well and free of disease after 10 years of follow up (Fig. 1c).

Pathologic Examination (Fig. 3)

Grossly tumour measured 5×3 cm. The tumor tissue was fixed in 10 % neutral buffered formalin, routinely processed and paraffin-embedded. Five-micron thick sections were cut for routine hematoxylin-eosin staining and

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Fig. 1 a Ulcerated tumour over scalp, b excised specimen, c photograph at 10 years of age

immunohistochemistry. For immunohistochemical staining, streptavidin biotin conjugate immunoperoxidase method was used, after appropriate antigen retrieval wherever required. Monoclonal antibodies against smooth muscle actin(dil; 1:100), vimentin (dil 1:100), desmin(dil 1: 100), myogenin(dil 1:100), ALK(dil 1:100) and β catenin(dil 1:100). All antibodies were obtained from M/S Dakopatts, Denmark. Multiple sections examined show a spindle cell lesion arranged in fascicles, at places showing herringbone pattern. Mitoses were seen but no necrosis was present. There was lymphocytic sprinkling amidst the tumour cells. The tumour cells were immunoreactive for vimentin and focally for smooth muscle actinin but were negative for myogenin, desmin, ALK and β

catenin (Fig. 3). Based on above features the diagnosis of infantile fibrosarcoma was made.

Discussion

Prenatal tumors are very rare, occurring with an incidence of 7.2 per 100,000 live births. Soft tissue tumors account for 25 % of congenital tumors and one third of these soft tissue tumors are malignant. Infantile fibrosarcoma (IFS) is one such malignant congenital tumor [8, 9]. Term IFS is used for fibrosarcomas occurring in children less than 5 years of age [1, 10]. However, IFS of scalp are extremely rare with only a few cases reported in the literature [2–7].

IFS are typically large tumors that grow very rapidly. These tumors are highly vascularised and may have ulcerations and bleeding, making it clinically difficult to differentiate them from hemangiomas [5]. IFS are more commonly seen in boys than girls and typically involve distal extremities (66 %) and trunk (25 %) [1, 7].

Despite histopathological similarities, IFS behave very differently to their adult counterpart. IFS has lower metastatic potential and better outcome as compared with fibrosarcomas occurring in adults [1, 7, 10]. Though, the local recurrence rate of IFS has been reported to be 20–50 % in literature. The rate of metastasis is low as compared to adults however it has been seen in up to 7–14 % [1, 6–8]. The role of adjuvant radiotherapy and chemotherapy is still controversial. There are some case reports of long survival without adjuvant therapy, however recurrences and distant metastasis have been reported even after adjuvant therapy [7–9].

RT-PCR can be used as a diagnostic tool in differentiating IFS from the adult fibrosarcoma by detecting the ETV6-NTRK3 fusion transcripts. A reciprocal translocation t (12;15)(p13;q25) that results in a fusion gene, ETV6-

Fig. 2 a MRI T1W1 axial cut, b MRI T1W1 Sagittal cut showing tumour over the scalp

Fig. 3 **a, b** (H & E) staining show a spindle cell lesion arranged in fascicles, at places showing herringbone pattern, **c** Mitoses were seen but no necrosis was present, **d** The tumour cells were immunoreactive for vimentin and focally for smooth muscle actinin

NTRK3 (ETS variant gene 6; neurotrophic tyrosine kinase receptor type 3) has been detected in IFSs [10].

Such lesions can be sometimes confused with hemangioma. However hemangioma does not enlarge and ulcerate so rapidly. Pre op biopsy of such lesions can lead to profuse bleeding due to highly vascular nature and friable margins, where bleeding can not be stopped by ordinary measures. In infants such endeavours should be undertaken in operation theater with all necessary back up including anaesthetist and blood; which was undertaken in this case.

Surgery has traditionally been considered the treatment of choice for IFS. The radicality of first surgery, aimed at achieving histopathologically negative margins, is associated with best prognosis [2, 7–9]. If a negative histopathological margin is obtained, long term disease free survival can be achieved even without any adjuvant therapy as happened in our case. However the role of adjuvant therapy is controversial in such cases. In patients with IFS involving the limbs, however, the issue of loss of function after surgery assumes importance. In such patients, chemotherapy has the potential of decreasing the size of tumor, thereby obviating the need of extensive surgery and making function preserving surgery a possibility [9].

This case emphasizes the importance of radical excision with negative margins is an important determinant of long term disease free survival in infantile fibrosarcoma.

References

- Orbach D, Rey A, Cecchetto G, Oberlin O, Casanova M, Thebaud E et al (2010) Infantile fibrosarcoma: management based on the European experience. *J Clin Oncol* 28:318–323
- Arigbabu SO, Durosinmi-Etti FA, Oyeneyin JO, Ojikutu NA (1985) Primary fibrosarcoma of the scalp: a review of a ten-year record from Lagos, Nigeria. *Clin Radiol* 36:465–466
- Tarik E, Lamiae R, Abdelouahed A, Tarik M, Hassan G, Anouar DM (2013) Unusual case of congenital / infantile fibrosarcoma in a new born. *Afr J Paediatr Surg* 10(2):185–187
- Kraneburg UM, Rinsky LA, Chisholm KM, Khosla RK (2013) Emergency surgical treatment of an ulcerative and hemorrhagic congenital/infantile fibrosarcoma of the lower leg: case report and literature review. *J Pediatr Orthop B* 22(3):228–232
- Jain D, Kohli K (2012) Congenital infantile fibrosarcoma: a clinical mimicker of hemangioma. *Cutis* 89(2):61–64
- Parkes SE, Muir KR, Southern L, Cameron AH, Darbyshire PJ, Stevens MC (1994) Neonatal tumours: a thirty-year population-based study. *Med Pediatr Oncol* 22:309–317
- Soule EH, Pritchard DJ (1977) Fibrosarcoma in infants and children. a review of 110 cases. *Cancer* 40:1711–1721
- Salloum E, Caillaud JM, Flamant F et al (1990) Poor prognosis infantile fibrosarcoma with pathologic features of malignant fibrous histiocytoma after local recurrence. *Med Pediatr Oncol* 18:295–298
- Kurkchubasche AG, Halvorson EG, Forman EN et al (2000) The role of preoperative chemotherapy in the treatment of infantile fibrosarcoma. *J Pediatr Surg* 35:880–883
- Sheng WQ, Hisaoka M, Okamoto S et al (2001) Congenital-infantile fibrosarcoma: a clinicopathologic study of ten cases and molecular detection of the ETV6-NTRK3 fusion transcripts using paraffinembedded tissues. *Am J Clin Pathol* 115:348–355