Locally Aggressive Orbital Fibrous Histiocytoma in A Child: A Case Report

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ABSTRACT

Fibrous histiocytoma is a pleomorphic sarcoma usually occurring in the deep soft tissue of old ages. It is rare in childhood. We report a 13-year-old girl with enlarged mass below the right eyebrow whose CT scan was in favor of soft tissue mass without bony erosion.

She was operated and histopathology was in favor of locally aggressive fibrous histiocytoma. Chemotherapy was given with VAC-D protocol for 12 months. Close follow-up 14 months after discontinuation of chemotherapy showed no sign of local recurrence or distant metastasis.

Key Words: Fibrous histiocytoma, Orbit, Childhood.

ÖZET

Bir Çocukta Lokal Agresif Orbital Fibröz Histiositoma: Bir Olgu Sunumu

Fibröz histiositoma, genellikle ileri yaşlarda derin yumuşak dokularda görülen pleomorfik bir sarkomdur. Çocukluk döneminde nadir görülür. Biz 13 yaşında bir kız çocuğunda sağ kaşının altında büyüyen bir kitle saptadık. CT'de kemik erozyonu olmayan yumuşak doku kitlesi saptandı.

Çocuk ameliyat edildi ve histopatolojik olarak lokal agresif fibröz histiositoma olduğu saptandı. Kemoterapi olarak VAC-D protokolü 12 ay verildi. Kemoterapinin kesilmesinden sonra 14 ay boyunca yapılan yakın takip sonucu lokal rekürrens ya da uzak metastaz saptanmadı.

Anahtar Kelimeler: Fibröz histiositoma, Orbita, Childhood

INTRODUCTION

Fibrous histiocytoma, a soft tissue tumor also referred to as fibrous xanthoma, was first described by Kauffman and Stout in 1961 (1). It is one of the diverse groups of soft tissue tumors usually occurring in deep soft tissue of later life (2). They account for one percent of all ocular masses. Ocular sign and symptoms vary, including decreased visual acuity, proptosis, diplopia, restricted extraocular muscle movement and swelling of the eyelids and conjunctiva (3). Fibrous histiocytoma is the most common mesenchymal neoplasm of the orbit in adults. Three types are recognized: benign, locally invasive and malignant subtypes (3,4). There are a few reports of malignant fibrous histiocytoma (MFH) of the orbit after radiotherapy for retinoblastoma (5) or pituitary adenoma (6). Fibrous histiocytoma is rare in children and its presence in the orbit is uncommon (7). The interest in this case relates to young age of the patient and rare site of the tumor.

CASE REPORT

A 13-year old girl was presented by her parents to the ophthalmologist due to an enlarged mass just below the right eyebrow. The tumor had been first discovered about 4 months previously and had enlarged slowly since then. The patient noted no tenderness when pressure was applied over the area. Ocular examination revealed a visual acuity of 20/20 in each eye and other examinations of the eye and adnexa were otherwise unremarkable. External examination revealed a horizontally oriented 20-10 mm firm subcutaneous mass in the supranasal area of the right orbit. A clinical diagnosis of dermoid cyst was made. Chest X-ray and all laboratory tests were normal. CT scan of the orbit revealed no bony erosion.

The patient underwent excisional biopsy of the lesion of the right anterior orbit. This was done under general anesthesia via an incision along the upper inner orbital margin. The excision was not complete since the tumor was not encapsulated and was adherent to the surrounding tissue. It was anterior to the septum. Histopathological examination showed admixture of fibroblasts and histiocytes (some forming giant cells) in storiform configuration (Fig. 1), (Fig. 2), (Fig. 3). Tumor cells invaded the neighboring striated muscles but frequent mitosis was absent (Fig. 4), so the diagnosis of locally aggressive fibrous histiocytoma was made.

The patient was referred to pediatric oncologist and chemotherapy was started with VAC-D protocol (Vincristin, Cyclophosphamide and Adriamycin alternated with Vincristin, Cyclosphamide and Actinomycin-D every 4 weeks for 6 months). Then, Etoposide was substituted for Adriamycin for another 6 months. Close follow up with CBC, ESR, physical exam monthly, chest x-ray, and orbital sonography every 3 months was done. Now she is well 26 months following the surgery and 14 months after discontinuation of the chemotherapy.

DISCUSSION

The ocular fibrous histiocytoma is a neoplasm composed of both fibrocytic and histiocytic elements. Raney et al (8) reported seven children aged 6 months to 11 years, three of whom had primary head and neck tumor. In a large series of 150 orbital fibrous histiocytoma reported by Font and Hidayat (4) the median age at presentation was 43 years and the youngest patient was 4 years of age. Based on the histopathologic features, the tumors are classified into three groups: benign, locally invasive and malignant. The rate of the recurrence was 31% for benign, 57% for the locally aggressive tumors and 64% for malignant tumors (4). Ulloa and Anderson (3) in a literature review stated that fibrous histiocytoma (either benign or malignant) account for one percent of all ocular masses and are the most common mesenchymal tumor of the orbit.

The lesion has also been reported in other sites including corneoscleral limbus and conjunctiva (9), subconjunctiva and anterior orbit (10), lacrimal sac (11), and tarsus (12). The lesions typically are difficult to remove in their entirety because of their infiltrative nature and there is a pronounced tendency for recurrence (11). In 150 cases of fibrous histiocytomas of the orbit who were reported by Font and Hidayat (4), 94 cases (62%) were benign, 39 cases (26%) were locally aggressive and 17 cases (11%) were malignant. The most common signs and symptoms were proptosis (60%), mass (46%) and decreased vision (25%) (4).

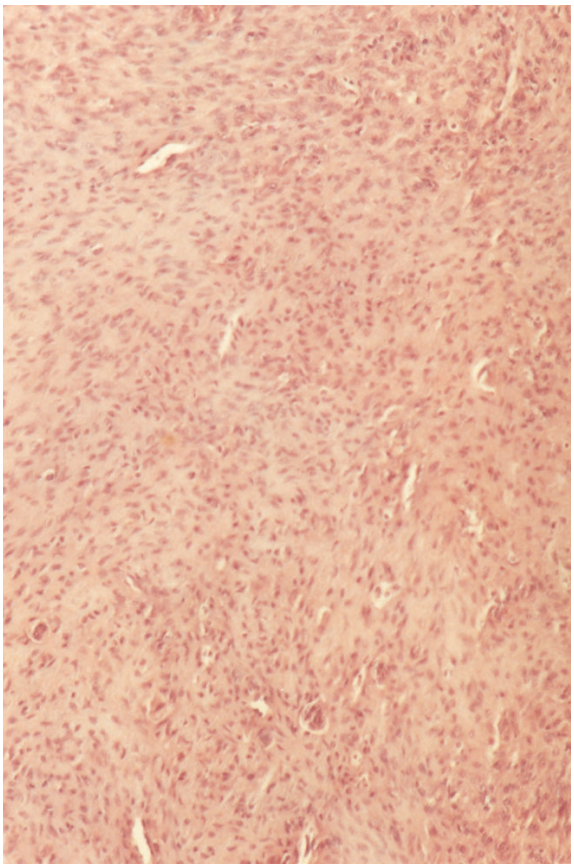

Figure 1. Tumor showing admixture of fibroblast and histiocytes in storiform configuration (H&E x 160)

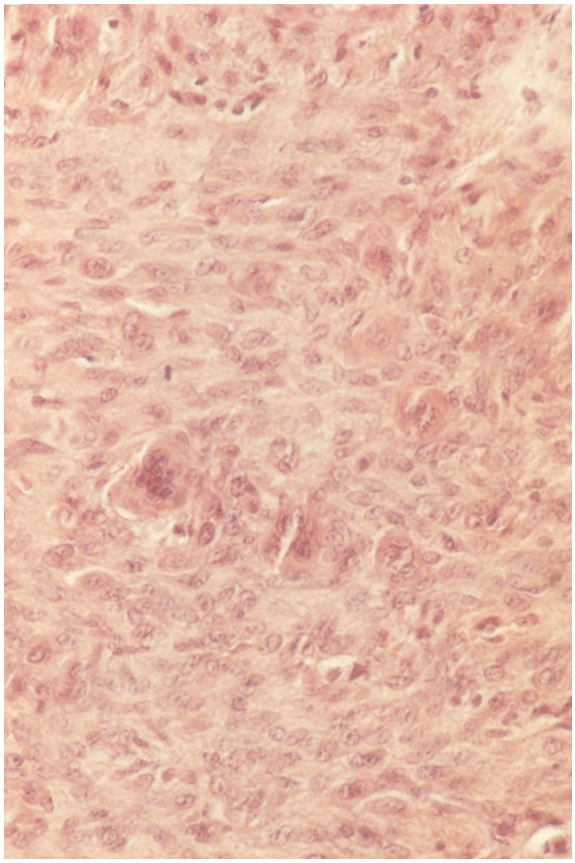

Figure 2. Fascicular arrangement of tumor cells with frequent giant cell formation (H&E x 220)

Although radiotherapy has been used in adult cases but due to the paucity of this tumor in children there is no report of large controlled trial on adjuvant radiotherapy in children (13). For our case; after group discussion in a tumor clinic (which was consisted from three hematopathologists, four radiotherapist-oncologists, three adult hematologists and three pediatric hematologist-oncologists) due to young age of the patient and good response to chemotherapy, it was decided to postpone radiotherapy, and prescribe it only if local relapse occurred. Combination of Vincristin, Cyclophosphamide with Adriamycin with or without Actinomycin-D, DTIC, Etoposide, Cisplatin which had excellent results in rhabdomyosarcoma has been suggested for other soft tissue sarcomas (8), and some authors suggested the use of Ifosphamide instead of Cyclophosphamide (13) .

Distant metastasis has been noted in 41% of 200 patients with malignant fibrous histiocytoma in 23 years collection of data (2) and the lung was the most common site of metastasis (82%). Bone, liver and regional lymph node metastases were also noted. Depending on the series reported by Daw et al (14), 18% of the patients with MFH had metastatic disease at the time of diagnosis. The only site of metastasis in this series was the lung whereas the lung, bone and regional lymph nodes were sites of metastasis in the study by Raney et al. (8)Favorable prognostic features were small primary tumor size (under 10 cm), superficial location, and possibly the presence of a prominent inflammatory response. Local recurrence rate was 41%.(8) In a series reported by Jakobiec et al (15), all the six cases of orbital fibrous histiocytoma had recurrence in 2-5 years after operation with marked tendency

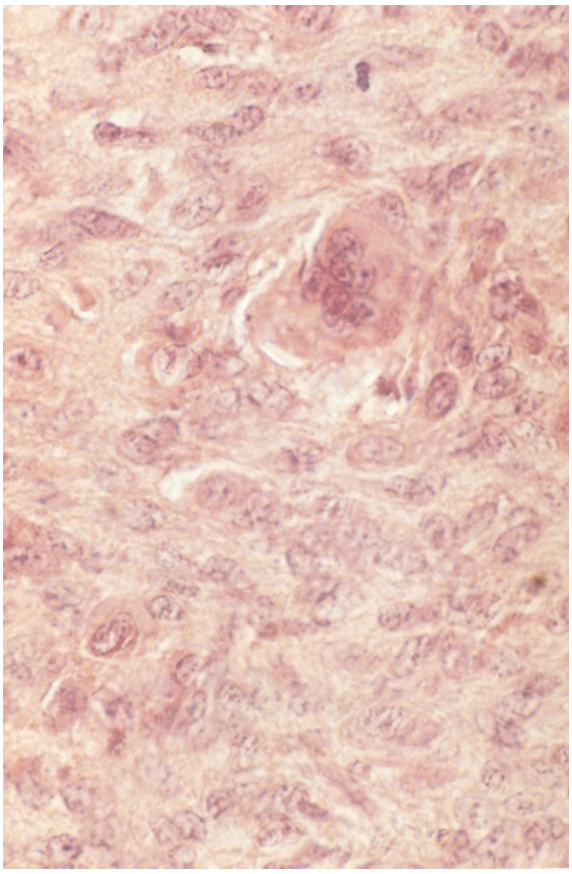

Figure 3. Higher magnification of the tumor cells showing the details of the giant cells and the tumor cells with ovid elongated vesicular nuclei and lack of mitosis (H&E x 460)

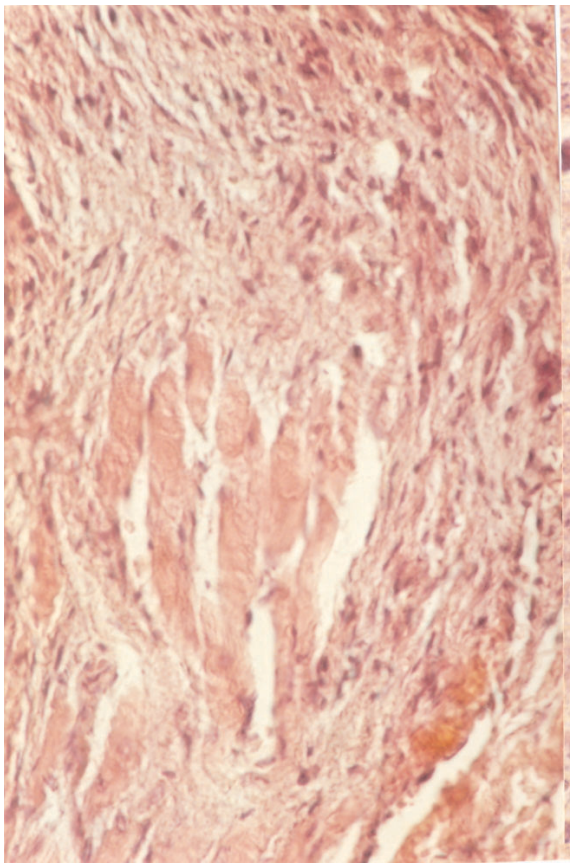

Figure 4. The tumor cells infiltrating and surrounding the adjacent skeletal muscle tissue (H&E x 220)

to invade fat, muscle, periostium or nerves. They believed that even the non-malignant fibrous histiocytoma had a tendency for metastasis. There is also a report of direct intraocular invasion of orbital MFH (16).

Our case had invaded surrounding skeletal muscle and did not possess the capsule. She fortunately had no distant metastasis at diagnosis and in the follow-up. Close follow-up for more than 10 years is needed because some local recurrence or distant metastasis may occur very late. (2) Ten year survivals of 17 patients of orbital MFH were reported to be 23% by Font and Hidayat. (4)

CONCLUSION

Orbital fibrous histiocytoma are rare periocular tumors that can manifest multiple ocular signs and symptoms. Even non-malignant fibrous histiocytoma have a high rate of recurrence, so careful histologic examination is necessary for diagnosis.

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