PRIMARY ORBITAL LIPOSARCOMA: Report of A Case

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ÖZET

Liposarkom, en sık görülen malign tümörlerden olmasına karşın, baş-boyun bölgesinde, özellikle de orbital kavitede oldukça nadirdir. Bu makalede, literatürdeki en genç vakalardan biri olan, yanlış klinik tanı ve yetersiz histopatolojik değerlendirme nedeniyle tedavisi geciken 14 yaşında bir kızda rastladığımız orbital liposarkom sunulmuştur.

SUMMARY

Liposarcoma, which is one of the most common malignancies of the body becomes one of the rarest ones when seen in the orbital cavity. An additional case of myxoid liposarcoma of the orbit in a 14 -year-old girl who is one of the youngest cases of the literature was presented. Because of clinical misdiagnosis and insufficient histopathological evaluation, the long course of the disease was also introduced in addition to the final surgical therapy.

Key Words : *Liposarcoma, orbital tumor, forehead flap.*

Since adipose tissue constitutes 15-25% of the body weight, liposarcoma is one of the most common soft tissue malignancies of the body, accounting for 16-18 % of sarcomas[1]. It was first described by Virchow in 1857[2]. Most frequent sites for liposarcoma are retroperitoneum and thigh, while only less than 4% of them occur in the head and neck region [3]. Among head and neck region liposarcomas, the most common site of occurrence is the orbital cavity and there are only 18 reported cases in detail, up to date[4,5,6]. This article aims to report an additional rare case of orbital liposarcoma and to discuss the results within the guidance of literature.

CASE REPORT

In July of 1993, a 14-year-old girl presented with a fragile, ulcerated mass protruding from the right orbital cavity which was also extending over nasal, maxillary and frontal regions (Fig.-1). She had a long medical history. She had first applied to Opthalmology Department in August. 1989 with a complaint of slight swelling on the right lower eyelid. The diagnosis was "inflammatory pseudotumor". After systemic and intralesionary corticosteroid therapy, she had been discharged as her symptoms subsided. In 1991, she had again referred to the same department with edema on upper and lower right eyelids and slight proptosis of the bulbus. At that time, right orbitotomy had been performed and the pathological diagnosis was "benign myxoid tumor", After recurrence of the symptoms in 1992, she had been operated in the same department and subtotal exenteration had been performed. Pathological specimen was classified as "myxoid liposarcoma". During postoperative follow-up in February. 1993 she had again been diagnosed as a recurrent sarcoma. She had been referred to Radiotherapy Department and a course of 6000 cGy was performed. According to the statements of the patient, a limited success was obtained and she was referred to our department as described above.

Physical examination revealed an 11x9x7 cm, easy bruising, nonpulsatile, nontender, round and lobulated firm mass. There was no

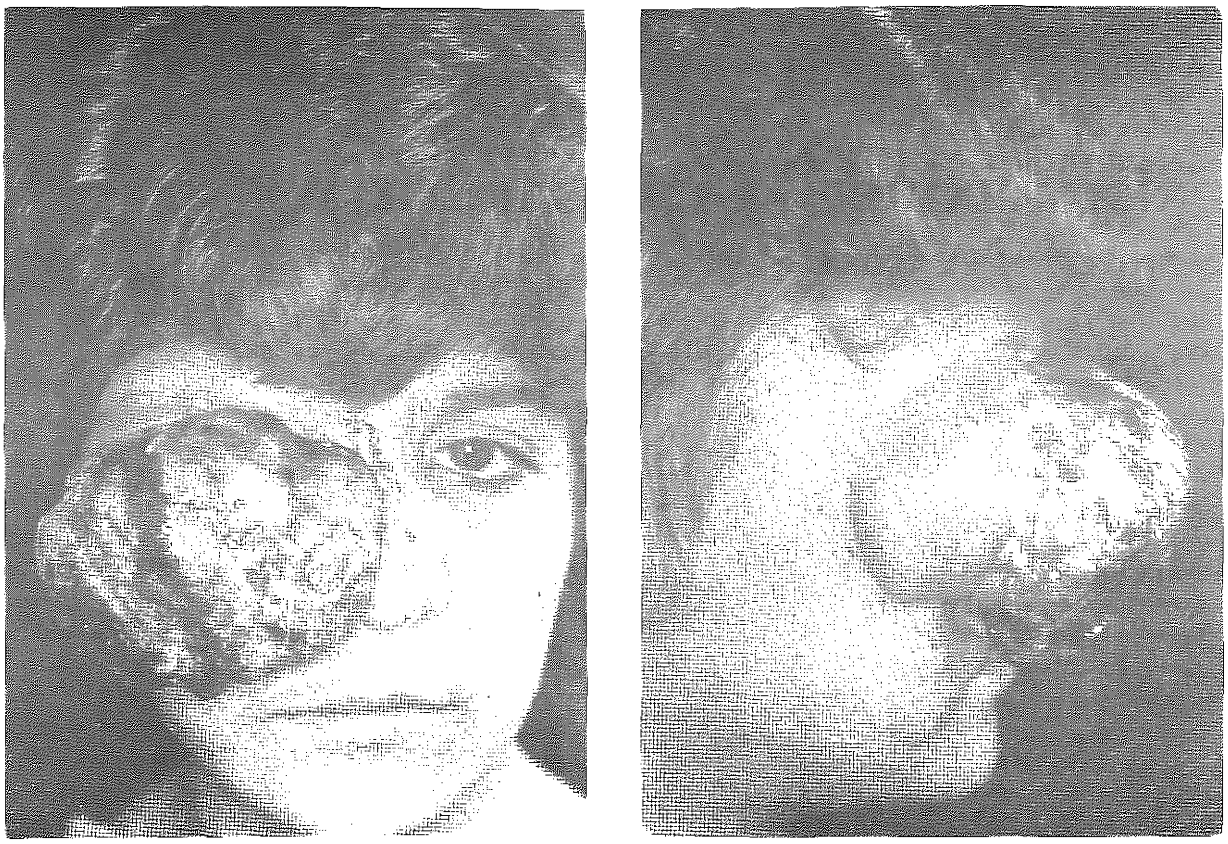

Figure 1: Easy bruising, petechial, round and lobulated mass of the myxoid liposarcoma protruding from the orbital cavity Preoperative frontal and lateral views.

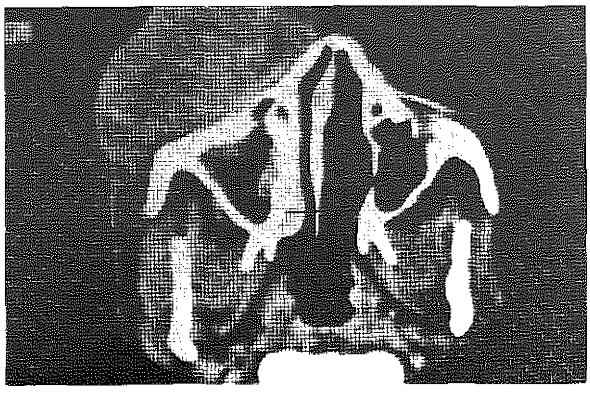

Figure 2: Sagittal CT scan showed no additional finding other than the mass affect of the tumor.

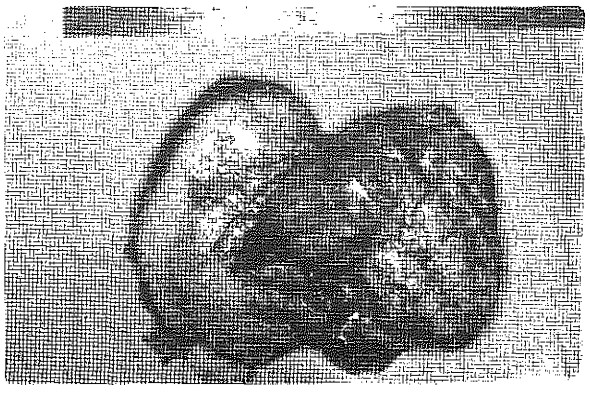

Figure 3: The 11 x 9 x 7 cm tumor, after resection.

palpable lymphadenopathy on the neck. In CT scan which has been taken in sagittal plane, there was no additional finding other than the protruding low density mass (Fig.-2).

The mass was removed with an excision passing through 2 cm below the lower eyelid, 1 cm medial to inner canthus and 5 cm lateral to external canthus (Fig.-3). Periosteum of the orbital cavity was included to the specimen. Lateral wall of the orbit was also eroded. After

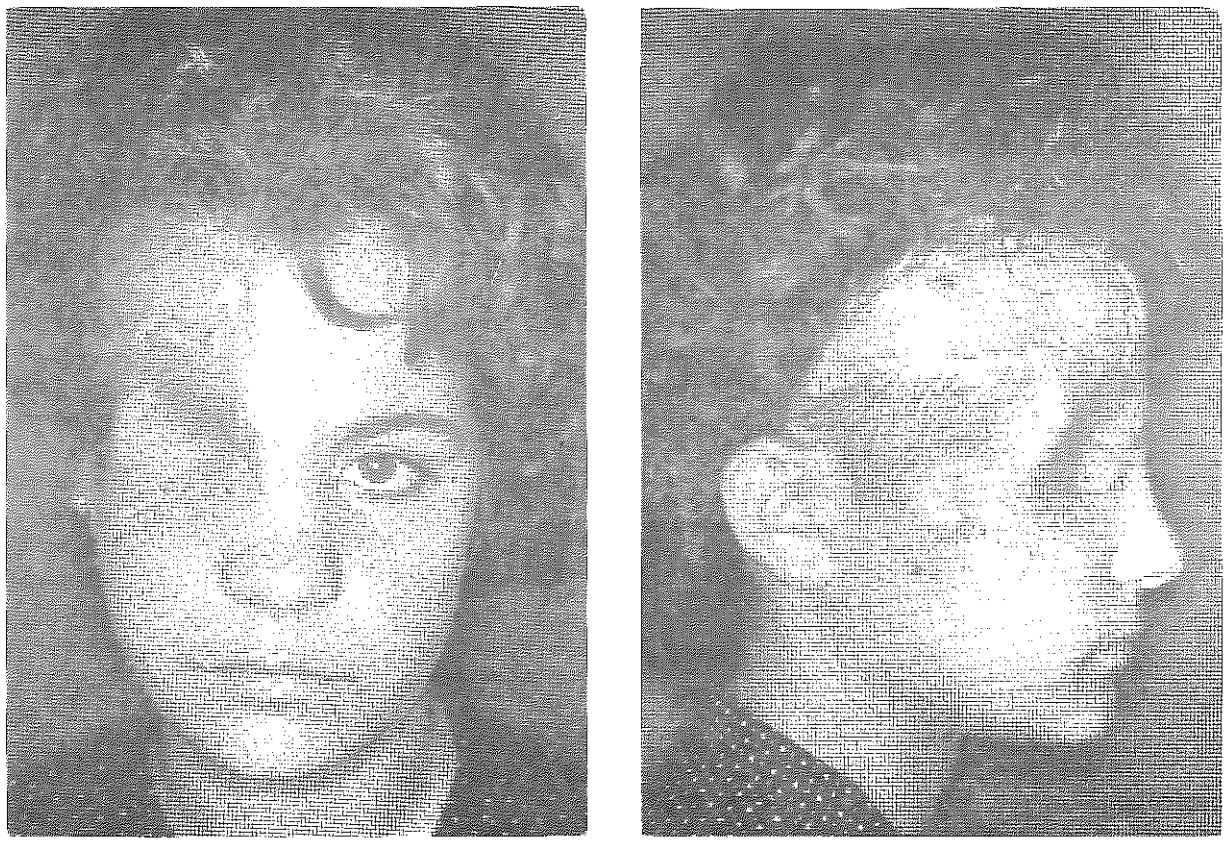

Figure 4: The defect was reconstructed with a pedicled frontal flap after resection of the tumor. Frontal and lateral views of the patient 2 weeks after surgery.

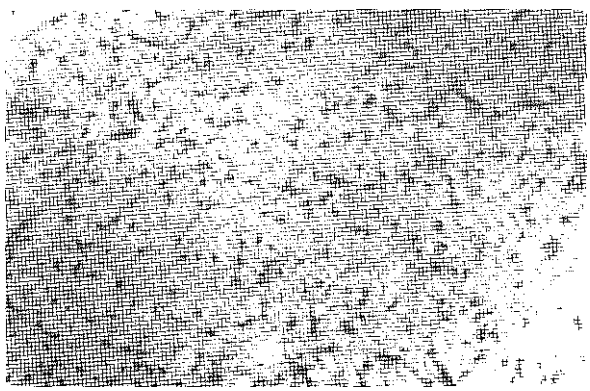

Figure 5: Atypical mesenchymal cells, mostly lipoblasts are seen in myxoid stroma (hematoxylin-eosin;original magnification, x200)

total resection of the mass, the defect was reconstructed with a frontal flap (Fig.-4).

The microscopic examination showed an extensive myxoid stroma containing preadypocytes with cytoplasmic vacuoles, collapsed vessels, rare mature adipocytes and mostly immature lipoblasts. There were also multinucleated giant cells, some hemorrhagic foci and necrotic spaces. The final diagnosis of Pathology Department was "myxoid liposarcoma" (Fig.-5).

DISCUSSION

The most common type of liposarcoma is the myxoid type and accounts for nearly the half of all liposarcomas. It has a favorable prognosis when compared to other subgroups. In myxoid liposarcoma there are spindle, stellate, uni- or multivacuolar lipoblasts suspended in a mucinous matrix. There may also be a plexiform capillary pattern background. Presence of mitotic activity or necrosis worsens the prognosis[1,7,8,9]. In our case, there were also a collapsed capillary background containing vacuolated lipoblasts

and also mature adipocytes. In some parts of the histopathological specimen there were multinucleated giant cells and necrotic spaces.

There are also cases of metastatic liposarcomas of the orbit in the literature and most common primary sites are retroperitoneum or thigh[1,10]. In order to rule out such an occasion, we have detected our patient with serial CT examinations.

While the peak incidence of liposarcomas are usually between the ages 40 and 60, orbital liposarcomas tend to be occur at a younger population. Males are slightly more commonly affected when compared with the females. At the time of first presentation, our case was 14 years old and she is the second youngest case in the literature. The youngest case is a 5-year-old child with an orbital liposarcoma reported by Quere et al.[11].

Common method for the treatment of liposarcoma is wide local excision, sometimes combined with radiotherapy. There is not a clear and documented approach for the use of radiotherapy yet. De Vita recommended radiotherapy in all liposarcomas at the postoperative period while some authors reserve radiotherapy only for nonresectable tumors or conservative therapy[12,13,14]. Our case was also consulted for radiotherapy at the end of second postoperative week and she was scheduled for a certain course.

Usually, liposarcomas grow by infiltration; poorly differentiated types commonly metastasize to lungs, but rarely to lymph nodes. Prognosis related to many factors such as histology, side, size and extent of excision[15].

Now, it is the fourth postoperative month of the patient and she is still under the control of Plastic Surgery, Radiotherapy and Medical Oncology Departments.

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