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Retroperitoneal Liposarcoma with Metastasis to both Orbits: an Unusual Metastatic Site

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Abstract. *Liposarcoma is among the most common adult histotypes of soft tissue sarcoma. It usually arises from the deep soft tissue of the extremities or the retroperitoneum. Despite the high content of fat in the orbit, both benign and malignant tumors of adipose tissue are extremely rare at this site; reports describing liposarcoma metastatic to the orbit are rare. Our report is a new case and the appropriate methods for diagnosis and treatment of this unusual metastatic site in this type of cancer are discussed.*

Liposarcoma is one of the most common soft tissue neoplasms, accounting for at least 10% of all sarcomas (1). It usually arises from the deep soft tissue of the extremities or the retroperitoneum.

Metastasis to the orbit from liposarcoma is rare, and only a few cases have been described in the literature (2-8). Our case of a man affected with retroperitoneal liposarcoma, who developed metastasis in both orbits and in the lung, is described and the diagnosis and treatment are discussed.

Case Report

A 53-year-old man had a low/intermediate-differentiated retroperitoneal liposarcoma diagnosed in April 2002, which was surgically removed. The patient was free of tumor recurrence for 2 years after its resection, when abdominal and lung metastases occurred. At that time, the patient started complaining of blurred vision and painless diplopia. Proptosis of both eyes became progressively evident. Extensive laboratory investigation including thyroid hormones and TSH was normal. Upon examination, visual acuity was 6/5 in the right eye and 6/4.5 in the left eye with limited extraocular movements in all directions. A computed tomography (CT scan) and magnetic resonance imaging (MRI) showed a bilateral retrobulbar mass that infiltrated the left lateral rectus muscle and the right inferior rectus muscle. In the MRI, the masses showed a hypointense signal intensity in the T1 sequences and a hyperintense signal in T2, with enhancement after Gadolinium-DTPA infusion (Figure 1, 2). The findings described were interpreted as metastatic bilateral orbital localization; 4 trans-septal left orbital biopsies were performed with local anesthesia, and metastatic tissue of liposarcoma was histologically confirmed. The patient was treated with external beam radiation therapy with a total dose of 30 Gy to the left orbit and high prednisolone doses were given. Thirty days after the end of radiotherapy, the patient received 6 courses of chemotherapy with Epirubicin 60 mg/m^2 days 1 and 2 plus Hyphosphamide 1500 mg/m^2 from days 1 to 5. The proptosis improved, but vision was permanently lost. Seven months after completion of chemotherap, the patient presented with progression of disease and started second-line chemotherapy.

Discussion

Liposarcoma is generally considered the most common soft tissue sarcoma and is mainly a tumor of adult life, with a peak incidence between 40 and 60 years of age and a slight male preponderance (1). The primary tumors occur most commonly in the thigh and retroperitoneum, while recurrence is common in deep-seated liposarcomas of all types, presumably because of the difficulty of complete surgical excision.

Metastases from liposarcoma are found mainly in the lungs, visceral organs, bone, sierosal surfaces of the pleura,

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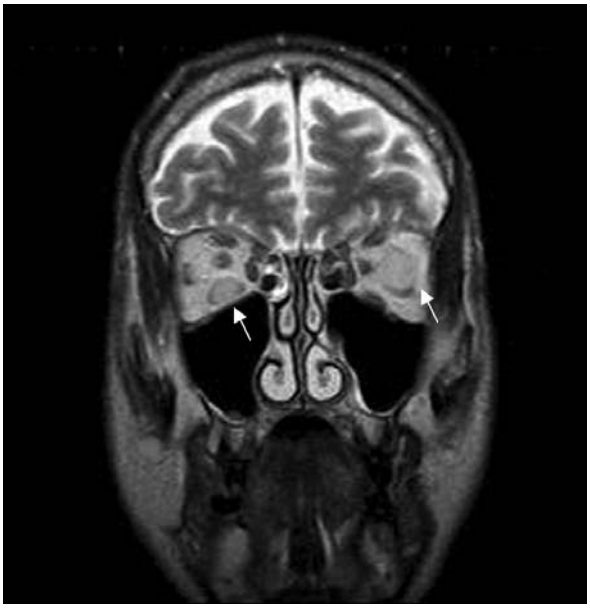

Figure 1. *T2-weighted image in coronal plane shows bilateral orbital masses (white arrows) with enlarged inferior rectus muscle on the right side.*

pericardium and diaphragm (1). Metastatic orbital liposarcomas are extremely rare, with only a few cases documented in the literature (1, 3, 8), so typical histological and imaging features can hardly be applied (4, 9, 10).

Almost every cancer can spread to the orbit. Carcinomas of the breast and the bronchial system are the most common sites of origin (5, 7). The diagnosis can be assumed on the basis of clinical and radiographic findings. In preseptal and parabulbar tumors, fine-needle aspiration biopsy (FNAB) is indicated (9). This diagnostic technique is swift and minimally invasive when in experienced hands. It can obviate the need for invasive surgery in patients in whom removal of the tumor is impossible without damaging the eye (5). In the case of concomitant orbital and visceral (lung) metastasis from liposarcoma, life expectancy is poor. When liposarcoma occurs in the orbit there is no specific pattern of involvement, and the tumor is characterized by marked diversity of clinical presentations.

The treatment of orbital metastases consists mainly of chemotherapy and radiotherapy, similar to the therapeutic

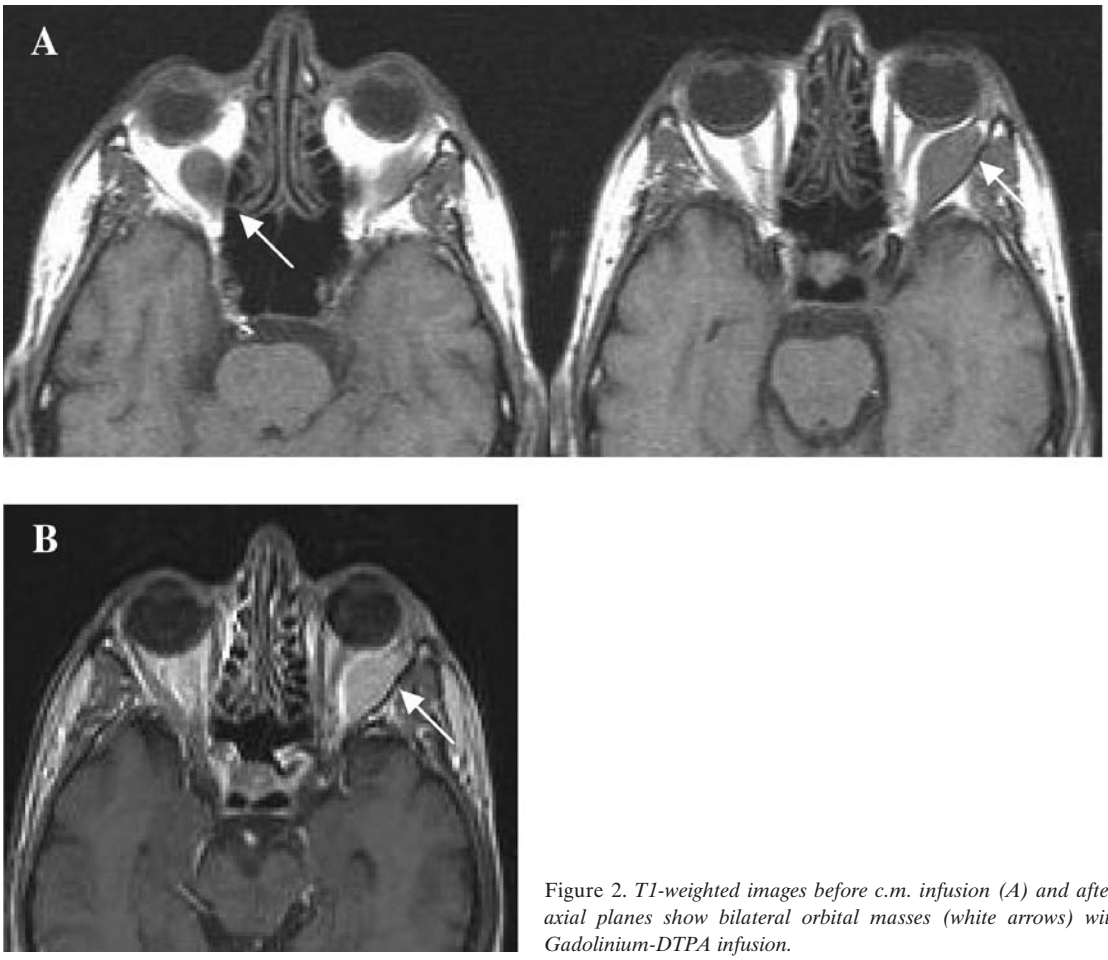

Figure 2. *T1-weighted images before c.m. infusion (A) and after c.m. infusion (B) in axial planes show bilateral orbital masses (white arrows) with enhancement after Gadolinium-DTPA infusion.*

management of metastases in other regions. Surgery is performed in order to obtain a histopathological confirmation of the diagnosis or to plan additional treatment. Surgery is also recommended if decompression by excision of the tumor is necessary. If the orbital mass is well circumscribed, excision may be suggested. This concept of treatment is similar to previously published suggestions (10). Aggressive surgical procedures, such as exenteration for cases of diffuse metastases infiltrating the orbit, should not be performed because the cosmetic effect is dramatic and the increase in survival time is questionable. On the other hand, the surgeon may be forced to perform this procedure if the patient complains of intractable orbital pain.

In contrast to the patient reported by Fezza and Sinard (3), our patient presented a tumor involving both orbits. Although preferential involvement of extra-ocular muscles has been documented, none of the previously reported cases of metastatic orbital liposarcoma described the synchronous involvement of both orbits and extra-ocular muscles of both sides.

Although the differential diagnosis of extra-ocular muscle enlargement is extensive and includes Graves's orbitopathy, myositis, carotid cavernous fistula, lymphoma and metastatic tumours, our case shows that physicians should include liposarcoma in the differential diagnosis of patients with an orbital mass and extra-ocular muscle enlargement.

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