LETTER TO THE EDITOR

Primary orbital liposarcoma: A case report

Le liposarcome orbitaire primitif : à propos d’un cas

Introduction

Primary liposarcoma is a malignant soft tissue tumor, which usually originates from mature adipose tissue and is most common in thigh and retroperitoneum [1]. Although liposarcoma represents the most common soft tissue sarcoma in adulthood, primary and secondary liposarcoma of the orbit are rare [2,3]. In part because of the rarity of such tumors, definitive initial histopathologic diagnosis is often difficult [4]. We describe a new case of primary orbital liposarcoma.

Case report

A 40-year-old female, with no past medical history, presented with right axial progressive painless non-pulsatile and non-inflammatory proptosis associated with a reduction in visual acuity (Fig. 1). Her best corrected vision was 5/10 P2 in the right eye and 10/10 P2 in the left eye. Anterior segment examination was unremarkable with no relative afferent pupillary defect, intraocular pressure and eye fundus was normal bilaterally. Ocular motility was not restricted. General examination was normal. A Goldmann perimetry revealed constricted visual fields.

MRI (Fig. 2) revealed an expansive intraconal mass with well-defined borders involving the optic nerve. T1-weighted images showed a hyposignal mass, on T2 and after gadolinium contrast, the mass showed a hyper intense signal. There was no destruction of the bone wall.

As presence of compression signs, surgical excision was performed and revealed a mass of hard consistency with greasy-yellow surface. Histological examination revealed proliferating spindle-shaped and stellate cells with lipoblasts suspended within myxoid-rich matrix and rich vascular pattern. The appearances were typical of a myxoid liposarcoma (Fig. 3).

Few months later, she developed recurrent painful proptosis with pain when moving the globe. The MRI revealed an ill-defined extraconal mass that was displacing the eyeball inwards. T1-weighted showed a hyposignal mass, on T2 and after gadolinium contrast, the mass showed a hyper intense signal (Fig. 4). The diagnosis of recurrent liposarcoma was made and surgical revision with adjuvant radiotherapy was performed. A total dose of 60 Gy was administered at 18 Gy/session, and was well tolerated. She was followed up for 12 months. There was no evidence of recurrence. On the last follow up examination, there was no proptosis of right eye. Her visual acuity was the same result as pre-operation, intraocular pressure was normal bilaterally. Both optic discs were normal. There was no evidence of the radiation neither retinopathy nor radiation optic neuropathy.

Discussion

Liposarcoma is rare, it represents 10%–20% of all sarcomas. [2]. The first description of orbital liposarcoma by Strauss appeared in 1911 [5]. Despite the fat richness of the orbit, primitive liposarcoma is rare with only 40 cases reported in the literature contrary to secondary liposarcoma [5–7]. Orbital liposarcoma with metastasis are even rare with only 6 cases reported [8].

Liposarcoma is more frequent in men than in women and is detected at an average patient age of 53 years. Compared with non-orbital liposarcoma, orbital liposarcoma has been shown to have a predilection to occur in younger patients [6].

The development of an orbital liposarcoma is often accompanied by clinical signs secondary to compression of orbital structures, the main symptoms are proptosis (92%), diplopia (42%), reduction in visual acuity (29%) and ptosis [1,5,6]. The duration of the symptoms before presentation ranges from 2 weeks to 7 years [9]. Radiological investigations including CT and MRI are essential to visualize the tumor and evaluate its local extension.

The tumor originates from primitive stromal cells related to intermuscular fascial planes or from perivascular pluripotential mesenchymal cells [6]. World Health Organization (WHO) recognizes five histologic types of liposarcoma: well-differentiated, myxoid, round cell, pleomorphic and dedifferentiated [10].

Classification based on karyotyping divides liposarcoma into the following three main subtypes: well-differentiated and its subtypes (12q13–15), myxoid/round (12: 16 translocation), and pleomorphic with complex karyotyping [11].

The link between the various types of liposarcomas is the lipoblast [6]. Myxoid liposarcoma of the orbit is the most common histological type (55%) [5], the larger proportion of myxoid types in the orbit correlates well with liposarcoma found in the soft tissues in general, where it is thought to account for 40% to 50% of all liposarcoma subtypes [6].

The clinical differential diagnosis of orbital liposarcoma includes any orbital mass with non-distinctive radiologic appearance [6]. The histological differential diagnosis of orbital liposarcoma is depending on the histological type. The well-differentiated type may mimic normal adipose,
main differences are the presence of variation in fat cell size and atypical hyperchromatic cells. Lipoblastic meningiomas with signet-ring cells may mimic liposarcoma but the vacuolated appearance of a lipoblastic meningioma is more prominent than in a true liposarcoma [2]. Myxoid pleomorphic lipoma can also mimic liposarcoma but tumor circumscription, presence of ropy collagen bundles, and lack of lipoblasts advocate myxoid pleomorphic lipoma [6].

Most cases of myxoid liposarcoma are composed predominantly or at least focally of bland fusiform to ovoid cells in a myxoid stroma with a prominent plexiform capillary network and scattered signet-ring lipoblasts. However, the morphologic spectrum of these tumors is great, and it is relatively easy to be misled by unusual histologic variants, especially when a small biopsy specimen reveals an unusual morphologic variant of myxoid liposarcoma. The diagnosis of myxoid liposarcoma is confirmed by fluorescence in situ hybridization studies for DDIT3 (also known as CHOP) rearrangement [11].

Sometimes initial histologic diagnosis is uncertain and modern immunohistochemical techniques using antibodies to S-100 protein, CD34, smooth muscle actin, and desmin may increase diagnostic precision [6].

Currently, Immunohistochemistry (IHC) for MDM2 and CDK4 can be used to provide strong support in distinguishing well-differentiated liposarcoma from other adipocytic neoplasms. The evaluation of MDM2-CDK4 amplification using fluorescence in situ hybridization (FISH) or quantitative real-time polymerase chain reaction (Q-PCR) can be used to supplement IHC analysis when diagnosis of adipose tissue tumors is not possible based on clinical and histological information alone [11].

The prognosis depends on several factors: size and location, histological type and grade of differentiation; well-differentiated and myxoid liposarcomas have a relatively indolent course, whereas the round cell and pleomorphic variants usually behave aggressively with more distant metastasis. Deaths from soft tissue liposarcoma in general are often as a result of the local effects of tumors and the oft inability to achieve clear resection margins because of disease location [4,6].

Treatment of orbital liposarcoma remains controversial with some surgeons performing repeated local resections for recurrence and others performing an initial exenteration after histologic confirmation of disease [6]. Indeed, surgical excision with ample margins admittedly improves survival and local control [12,13], but most of the orbital liposarcomas cannot be completely excised without causing damage to infiltrated structures, particularly the extraocular muscles [6]. Association of radiotherapy provided best results.
Myxoid liposarcomas are believed to be radiosensitive if not radiocurable but well-differentiated tumors are less responsive [6]. Conservative surgery is indicated for small, well-defined and easily accessible tumors, with adjuvant radiotherapy, while exenteration is recommended for invasive, dedifferentiated and recurrent tumors [6,12]. The use of chemotherapy is controversial, it has been reported that dexorubicine and dacarbazine are effective in the treatment of myxoid liposarcoma [6,15].

Conclusion

The primary orbital liposarcoma is a rare entity; the positive diagnosis is based on imaging and confirmed by histological study after excision biopsy, diagnosis may be difficult because of initial histopathologic uncertainty. Exenteration after confirmatory biopsy seems to provide the best chance of a cure, with increasing evidence for the use of adjuvant radiotherapy. Prognosis and management depend on several factors including the size and histological type.

Disclosure of interest

The authors declare that they have no conflict of interest concerning this article.

References


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