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Case Report

A Rare Orbital Pathology: A Large Orbital Dermatofibrosarcoma Protuberans

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Abstract

Dermatofibrosarcoma protuberans (DFSP) is a rare sarcoma of the dermis. It is a malignant, locally aggressive, and infiltrative tumor with frequent recurrence. In this case, a 44-year-old woman presented with a 15-year history of a swelling in the medial canthus of the right eye that caused tearing. Imaging revealed a septated mass isodense to soft tissue that had eroded the medial wall of the orbit. Macroscopic examination showed an elastic, gray-brown, encapsulated, irregular mass measuring 45x35x22 mm. The surgical margins were positive, so adjuvant radiotherapy was started. The patient was followed for 2 years without recurrence. According to the literature, this mass is the largest orbital DFSP treated by globe-sparing primary resection.

Keywords: Dermatofibrosarcoma protuberans, orbital tumor, orbital radiotherapy, medial canthus tumor, eye tearing

Introduction

Dermatofibrosarcoma protuberans (DFSP) is a primary, well-differentiated mesenchymal tumor of the dermis with an incidence of 0.8-5 per million. It constitutes 0.1% of malignancies in the head and neck region and 1% of soft tissue sarcomas.[1] It is a locally aggressive tumor with a high rate of local recurrence and low metastasis rate.[2] Treatment consists of excision with a wide safety margin. In the presence of recurrence and metastases or in cases where surgical treatment cannot be performed, radiotherapy and chemotherapy are alternative methods. According to the literature, 14 cases of orbital DFSP have been reported, and the mass presented here is the largest DFSP treated by globe-sparing primary resection. It is also the first case of orbital DFSP reported in Turkey.

Case Report

A 44-year-old woman presented to our clinic with complaints of progressive swelling over the right medial canthus region for 15 years that was causing eye tearing. She had a history of a previously excised mass at the same location 20 years ago, with no pathology report. On ophthalmologic examination, best corrected visual acuities were 20/25 in the right eye and 20/20 in the left eye. A non-tender, rubbery, immobile mass was palpated over her right medial canthus (Figure 1A). The right globe was displaced laterally without any motility restriction. Anterior

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and posterior segment findings were unremarkable except for prominent tearing on the right.

The mass was visualized preoperatively with magnetic resonance imaging (MRI). A hypodense mass (isodense to soft tissue) was detected on T1-weighted MRI (Figure 2A). MRI showed heterogeneous contrast enhancement in T2-weighted and short tau inversion recovery sequences. There were septa inside the mass and the bones showed erosion and remodeling. It was observed that the mass originated from the extraconal space (Figure 2B), reached the ethmoidal cells, and breached the medial orbital wall (Figure 2C). There was destruction of the lamina papyracea and ethmoidal septa, with a 3.5-mm defect in the fovea ethmoidalis and a millimetric defect in the lateral lamina of the cribriform plate. The mass obliterated the nasolacrimal groove, displaced the globe laterally, and grew posteriorly, nearly reaching the brain (Figure 2D).

Excisional biopsy under general anesthesia was planned. Following a right Lynch incision (Figure 3A) and orbicularis dissection, the mass was visualized, carefully dissected (Figure 3B), and removed en-bloc (Figure 3C). Intraoperatively, a small orbital wall defect measuring 3x2 mm was seen in the lamina papyracea, but the underlying mucosa was intact. No tissue representing the lacrimal sac was detected in the orbit. After removing the mass, we inspected for cerebrospinal fluid leakage but did not perform any additional tests.

Macroscopic inspection of the specimen revealed an elastic, gray-brown, encapsulated, irregularly surfaced mass measuring 4.5x3.5x2.2 cm in size (Figure 3D). In the sections, the inner tissues were tan-pink colored with a softer consistency. The tumor cells were positive for vimentin (Figure 4A) and CD34 (Figure 4B) and negative for CD31, Factor 8, EMA, CD68, desmin, and S-100. The proliferation index was 5% (Ki-67) (Figure 4C). The tissue margins were positive at multiple locations because the tumor's proximity to the brain and globe made a wide margin excision impossible.

Due to the positive surgical margins, the patient was treated postoperatively with 5400 Gy (180 Gy/fraction/day) intensity-modulated radiation therapy applied using helical tomotherapy. The patient was followed for 2 years with no recurrence and no distant metastasis (Figure 1B). However, she did not come to follow-up visits after 2 years.

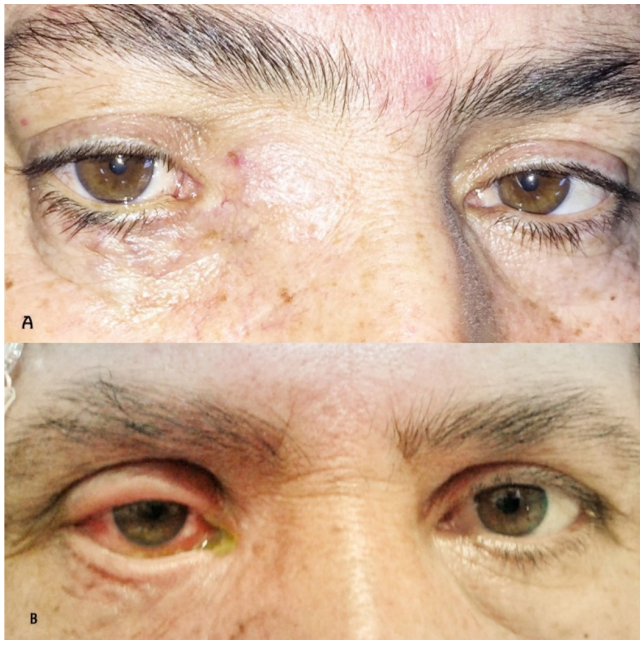

Figure 1. A) Preoperatively, swelling due to a mass completely under the skin is seen in the medial canthus region. B) Postoperative photo showing eyelash loss, eyelid edema, and conjunctival hyperemia due to radiation treatment. Persistent tearing is also seen, which is the result of both surgery and radiotherapy

Discussion

DFSP is seen most frequently between the ages of 20-50 years, with male predominance. DFSP occurs most often on the trunk and least commonly in the head and neck. Only 3.5% of DFSPs of the head and neck involve the periocular region.[1] Among orbital cases, half of patients are over 60 years of age and the gender ratio is equal. Orbital cases are most frequently in the medial canthus area, followed by the lower and upper eyelids.[3]

The tumor usually appears initially at a single focus, in the form of a mobile, pink, fibrotic skin nodule that does not adhere to deep tissues. Apart from the solitary nodule, there are also flat sclerotic and atrophic plaque forms.[4] It was named protuberans because of its protruding structure. It may occur in scar areas secondary to trauma or vaccination, in areas exposed to ionizing radiation, or spontaneously.

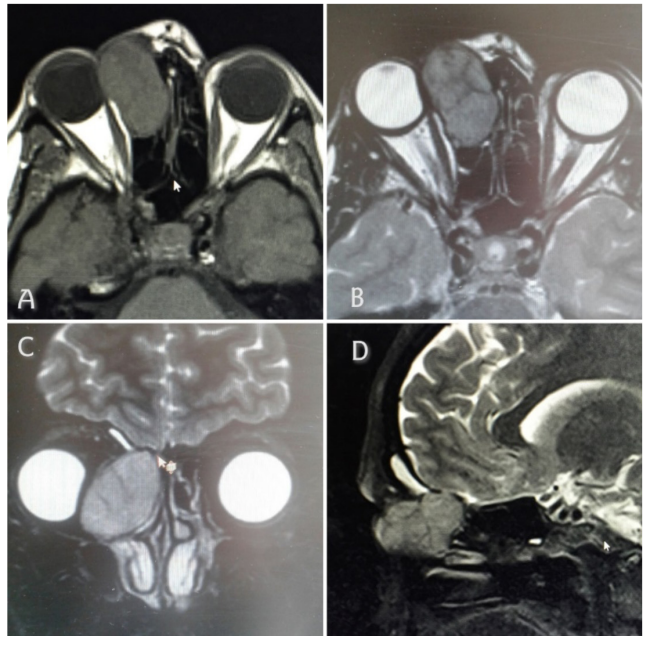

Figure 2. A) A hypodense mass (isodense to soft tissue) was detected on T1-weighted magnetic resonance imaging (MRI). B) In the axial section of the MRI, a septated mass originating from the extraconal space and causing destruction of the bones was observed. C) The coronal section of the MRI showed that the mass reached the ethmoid cells and pushed the medial orbital wall. D) The sagittal section of the MRI showed a 3.5-mm defect in the fovea ethmoidalis and the mass extending posteriorly and approaching the brain

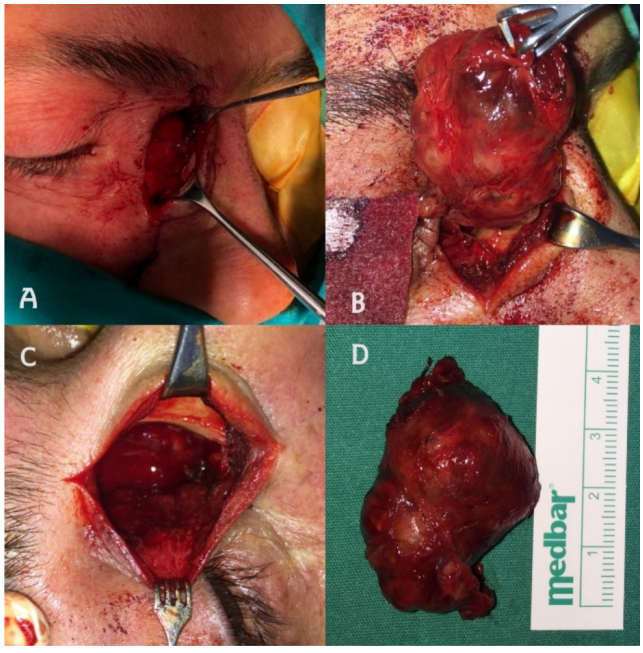

Figure 3. Intraoperative images of the right Lynch incision (A), the dissection (B), the mass removed en-bloc (C), and the irregularly surfaced mass measuring 4.5x3.5x2.2 cm in size (D)

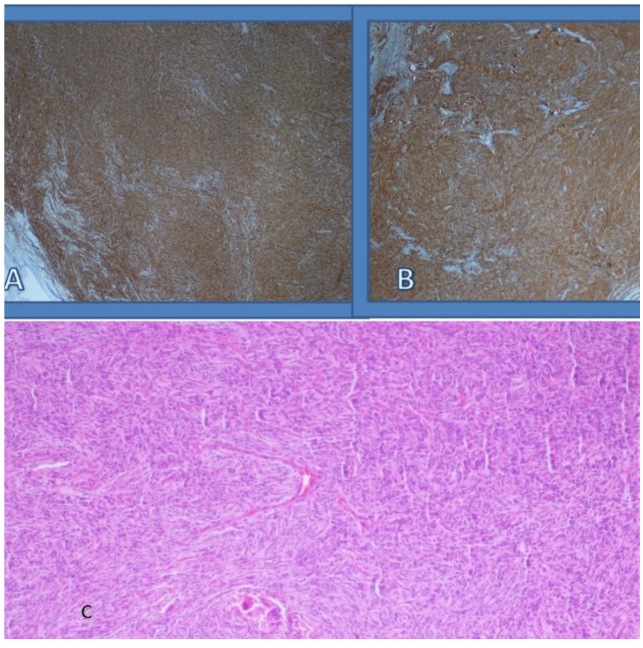

Figure 4. Diffuse strong vimentin (A) and CD34 (B) staining (x40) and vortex-like array of monotonous spindle tumor cells (C) (hematoxylin and eosin, x100)

It tends to grow by infiltrating the surrounding skin, subcutaneous, muscle, and bone tissues. Therefore, MRI and computed tomography are very useful in demonstrating local involvement during diagnosis, follow-up, and treatment.

The tumorigenesis of DFSP involves the t(17;22)(q22:13) reciprocal translocation.[5] The expression of CD34 and vimentin strongly supports the diagnosis of DFSP. Some tumors may be SMA-positive in focal areas, but S100 positivity is not observed. In our case, CD34 and vimentin were positive, while SMA and S100 were negative.

The main principle in the treatment of DFSP is excision with a wide margin of safety. Frozen section analysis and Mohs surgery are controversial because of the high likelihood of local recurrence.[6] Low-grade, cutaneous sarcoma with autocrine overproduction of the platelet-derived growth factor (PDGF The dense connective tissue found at the periphery of the tumor appears as a pseudocapsule that has irregular finger-like cellular protrusions invading surrounding tissues. As this tumor is mostly seen in the trunk and extremities, removing 3-5 cm with normal tissue is recommended in those exposed areas to prevent recurrences. However, this is impossible in the orbital area. Local recurrence is around 20-50% and mostly occurs within three years. The earliest local recurrence reported for orbital DFSPs was observed 1 week after excision.[7] In a previous case of orbital DFSP, exenteration was performed in addition to tumor excision due to the detection of positive surgical margins.[8] The 5-year survival rate is reported to be 93-100% for excisions made with adequate safety margins.[2]

Radiotherapy and transforming growth factor-beta inhibitors are other promising treatment options for DFSP, but their routine use is still controversial.[9] Adjuvant radiotherapy was reported to improve 10-year local control and survival rates in cases with positive surgical margins, local invasion, and recurrence.[9] Therefore, adjuvant radiotherapy and chemotherapy should also be kept in mind to avoid exenteration. These patients should be followed up for life, as local recurrence can be seen after many years.

Ethics

Informed Consent: Obtained.

Peer-review: Externally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: A.A.K., Z.G.Ö., A.G., M.N., Concept: Z.G.Ö., Design: Z.G.Ö., Data Collection or Processing: A.A.K., Z.G.Ö., A.G., M.N., Analysis or Interpretation: Z.G.Ö., A.G., Literature Search: A.A.K., M.N., Writing: A.A.K., Z.G.Ö.

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