Recurrent chordoma with orbital and eyelid invasion

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Key Words: Chordoma, orbital invasion, diplopia, proptosis, cranial nerve palsy.Dear Editor,

Herein we present an interesting and rare case of aggressive orbital and eyelid chordoma with imaging, morphological and immunohistochemical confirmation.

**A 51-year-old man was referred to ophthalmology with right-sided enophthalmos and hypoglobus with diplopia following surgery for his skull based chordoma. The patient had a complex 6-year history of recurrent chordoma affecting the odontoid peg treated with repeated surgical debulking and radiotherapy (he was deemed unsuitable for proton beam). His most recent surgery involved a right lateral rhinotomy and maxillectomy after which he was left with enophthalmos, hypoglobus and presenting complaint of intermittent diplopia. He was also under the care of oral and maxillofacial unit for speech and swallowing difficulties. On examination the patient had an abnormal head posture with a right head tilt and face turn to the left to help overcome his moderate right hypotropia and small exotropia (Fig.1a). He had right infraorbital numbness and reduced right corneal sensation. Vision was 6/5 in both eyes with normal colour vision and pupillary responses, otherwise normal anterior segments and fundoscopy. The patient underwent a right orbital floor repair with a MEDPOR TITANTM implant (Stryker). Following surgery, his diplopia improved, the eye was in a better position (Fig.1b) with good binocular motility and vision. However, 6 months post-orbital surgery small, firm, immobile subcutaneous lumps were noticed on the right lower lid, over lateral canthal area and lateral orbital rim. Computerised tomography revealed a right supraorbital rim and temporal fossa mass and hence the patient was listed for biopsies of the right lower lid/lateral canthal and orbital lesions (histology below).**

**Interestingly, during induction of the general anaesthetic prior his orbital/lid surgery, the patient was noted to have a mass in the neck. Subsequent** magnetic resonance imaging **of the neck revealed likely** recurrence of the chordoma in the area of previous decompression with clival involvement. The patient is awaiting further debulking surgery.

**Histological examination of all the tissue fragments from the lid and orbital biopsies showed a myxomatous tumour composed of cords, trabeculae and fibrous stroma surrounding nests of atypical physaliferous cells with eosinophilic partially vacuolated cytoplasm and varying-sized nuclei (Fig.2a&b). On immunohistochemistry (IHC), the atypical cells were positive for** epithelial-membrane-antigen **(Fig.2c), pancytokeratin marker MNF116 (Fig.2d), cytokeratin 19 (Fig.2e), vimentin (Fig.2f), brachyury (a specific marker for chordoma; Fig.2g) and negative for tumour suppressor p27Kip1, D2-40 and glial-fibrillary-acid-protein. Ki-67 growth-fraction was <2%. Morphological and IHC analysis confirmed the diagnosis of chordoma.**

Chordomas are slowly growing embryonic tumours derived from the notochord vestiges which occur along the axial skeleton. Current management involves combining maximal surgical resection with high-dose proton radiation therapy, adjuvant chemotherapy does not improve survival [1-2]. Reports of orbital chordoma are extremely rare and usually limited to single case reports, with only 21 cases being reported between 1937 and 2016. Evaluation of treatment outcomes and survival of chordoma is thus difficult, because of their rarity and the lack of follow-up data. The few cases reporting orbital involvement have shown strong recurrences [3-5]. A possible explanation may be seeding during debulking or surgical resection of the tumour. The ten year recurrence-free survival has been reported to be between 31-39% for skull base chordomas but 26% in reoperated cases [6-7]. Little is known of survival rate of patients with orbital chordoma, and such information may have to be extrapolated from the clinical course of patients with skull base chordomas.

Herein we present a case of aggressive chordoma where interestingly the latest recurrence was detected at the time of anaesthetic on day of surgery for suspicious peri-and orbital lesions. These lesions were all histologically confirmed chordomas. To the best of our knowledge, this is the first case report with local invasion into the eyelid. Previously there has only been one case with metastatic chordoma to the eyelid reported in the literature [8]. Although VIth cranial nerve palsy has been reported as a first sign of clival chordoma [4]our patient presented with diplopia from a right exotropia and hypotrophia following lateral rhinotomy with maxillectomy for his skull base chordoma. This improved with post-orbital floor repair.

In conclusion, this case demonstrates recurrent chordoma with orbital and eyelid invasion with imaging, morphological and immunohistochemical confirmation. Interestingly, optic nerve function was preserved and other orbital signs only developed following extensive previous surgery but improved post orbital floor repair unlike those reported previously.

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Ethical approval:- All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent:- Informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this article. The study was conducted in accordance with the declaration of the Declaration of Helsinki. The patient had consented to the submission of the letter to the journal.

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Fig. 1

a) Preoperative photograph showing hypoglobus with hypotropia and CT scan image of the patient (white arrow points to tumour mass in superotemporally within the orbit); b) Photograph and CT image taken of the same patient post right orbital floor repair. The orbital mass can still be seen in the superior aspect of the orbit (white arrow). The position of the orbital floor implant is also shown by the dashed arrow). Note position of pupillary reflexes in colour photographs pre- and postoperatively

Fig. 2

Tissue fragments of: a) Right lower lid lesion (H&E, x4 objective) and b) Orbital lesion (H&E, x10 objective) containing **a myxomatous tumour composed of cords, trabeculae and fibrous stroma surrounding nests of atypical physaliferous cells.**  These vacuolated tumour cells are seen invading the muscle (indicated by black arrow). IHC showing positive immunostaining (3,3'-Diaminobenzidine [DAB] x10 objective) for: c) epithelial-membrane-antigen**; d) pancytokeratin-marker MNF116; e) cytokeratin 19; f) vimentin and g) nuclear brachyury (x5** objective**); h) p27 Kip1 staining is negative (x5** objective**).**