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CASE REPORT

Recurrent inverted papilloma of paranasal sinus presenting as acute proptosis

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Abstract

Objective. To describe the course of events that followed from the time of the diagnosis to the management of a rare case of recurrent inverted papilloma presenting as an acute proptosis.

Methods. A seventy-year-old diabetic female patient presented with a painful left eye proptosis for 15 days. She had a history of resection of inverted papilloma of paranasal sinus followed by radiotherapy for eight years before. The examination revealed a 23 mm proptosis, with restricted ocular movements, corneal oedema, funnel shaped anterior chamber, and total retinal detachment with a complete visual loss. The lobulated fixed hard mass was palpable circumferentially but more in the inferior orbital compartment. The transconjunctival incisional biopsy showed features of highly undifferentiated cytology. The lid sparing exenteration was done under general anesthesia with cosmetic reconstruction.

Results. Immunohistochemistry of exenterated mass was doubtfully suggestive of a small cell tumor. However, histopathology confirmed features of rhabdomyosarcoma.

Conclusion. The present case study revealed rhabdomyosarcoma cytology presenting as an association-inverted papilloma.

Keywords: inverted papilloma (IP), paranasal sinus (PNS), squamous cell carcinoma (SCC), acute proptosis, exenteration, rhabdomyosarcoma

Abbreviations

IP = Inverted papilloma, PNS = Paranasal sinus, SCC = Squamous cell carcinoma, IOP = Intraocular pressure, CT = Computed tomography

Introduction

Inverted papilloma (IP) are usually benign, however, locally aggressive tumors of sinonasal cavity are known to recur after the initial tumor resection. They arise from the lateral wall of the nose, frontal and ethmoidal sinus, and lacrimal system. They have been found to have a malignant transformation in 5 (13%) cases [**1**]. Orbital metastasis was reported particularly for squamous cell carcinoma and transitional cell carcinomas [**2**]. Six out of ten patients with inverted papilloma showed a transformation to squamous cell carcinoma, four patients had a transformation to transitional cell carcinoma and eight patients had an orbital involvement that recurred after the initial resection [**3**].

The inverted sinonasal papilloma (Ringertz tumor) may present anywhere within the nose and paranasal sinuses, occasionally within the

upper aero digestive tract. The epithelium may be squamous, glandular, transitional cell-like or a mixture in any combination and permutation with synchronous or metachronous multicentric on the clinical presentation [**4**].

The present case study reported a rhabdomyosarcoma association with inverted papilloma, presenting as an acute unilateral painful proptosis and its management. Regarding the best literature search performed by us, this was the first case report of IP associated with rhabdomyosarcoma cytology.

Methods

A seventy-year-old diabetic female patient presented with an acute painful left eye proptosis that lasted for 15 days (**Fig. 1**). There was documentary evidence of an endoscopically assisted tumor resection that aroused from the medial wall of the left maxillary sinus and the frontal recess, performed eight years before, which was diagnosed on computed tomography (**Fig. 2**).

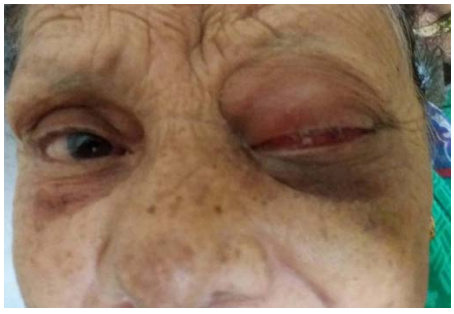

Fig. 1 Acute proptosis due to transorbital extension by recurred inverted papilloma on presentation

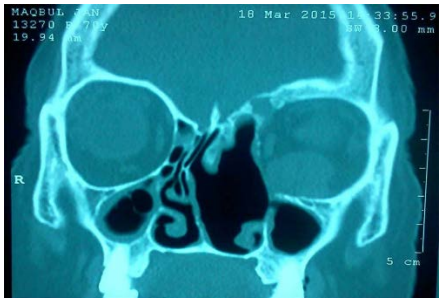

Fig. 2 Computed tomography showing previously treated inverted papilloma with recurred mass in the inferior orbital compartment

Transnasal endoscopic resection was done 8 years before for a benign inverted papilloma along with a medial maxillectomy followed by radiotherapy to prevent the recurrence as the excised mass revealed features of squamous cell carcinoma. Sinonasal squamous cell carcinoma features were revealed after the endoscopic resection of the benign inverted papilloma on histopathology. Histopathologically, the mass was confirmed as being malignant inverted papilloma with features suggestive of squamous cell carcinoma. Hence, the patient was advised to undergo post surgical radiotherapy. The patient had a recurrence free gap of 8 years, during which, she was not followed up.

While examined with a Hertels exophthalmometer, the left eye showed a proptosis of 23 mm. A tender, lobulated, fixed mass of firm to hard consistency was palpable circumferentially, more in the inferior orbital compartment. The inferior globe rotations were predominantly restricted and painful with limited up gaze movements. A provisional diagnosis of recurrence of inverted papilloma with direct orbital metastasis was considered clinically and on magnetic resonance imaging. The cornea demonstrated a uniform haziness and a funnel shaped anterior chamber with total ring posterior synechiae. The intraocular pressure (IOP) was 43 mm Hg by rebound tonometer with an absent glow on ophthalmoscope. The right eye was pseudophakic with a 6/ 6 vision with an unremarkable ophthalmoscopy.

The B scan of the left eye showed a funnel shaped retinal detachment with an orbital mass silhouette. Nasal endoscopy revealed previous surgically induced adhesions, a wide osteomeatal complex with intact and bulged out lamina papyracea. The computed tomography (CT) of the paranasal sinus (PNS) showed an irregular extraconal soft tissue mass lesion, abutting inferior rectus muscle with a previous post operative defect in the floor of the left frontal sinus and an anterior part of the lamina papyracea with a left temporal lobe showing white matter hypodensity. The MRI brain and orbit revealed previous postoperative changes in the left osteomeatal and fronto ethmoidal sinus. An ill-defined enhancing soft tissue mass lesion of size 25x22x19 mm involving the inferior rectus muscle at the floor of the left orbit which

was hypointense on T1 and hyperintense on T2 weighted images, was observed. Lacunar infarcts involving both the corona radiata and the centrum semiovale region with gliotic changes, probably due to radiotherapy were observed in the left temporal lobe.

A transconjunctival approach for biopsy was preferred to the transnasal route, as an opening lamina papyracea could predispose to the seedling of the tumor cells into the ocular adnexa and systemic circulation. As the inferior conjunctiva and the orbital septum were incised, a profound haemorrhage was observed and haemostasis was attained by bipolar cautery. Bits of tissue were subjected for histopathology and immunohistochemistry. There was an alleviation of pain on the first postoperative day, with some gain on the inferior globe movements.

The histopathological examination of the recurred mass revealed a highly undifferentiated cytology suggestive of squamous cell carcinoma and the immunohistochemistry reported a small cell tumor, possibly a carcinoid tumor (**Fig. 3**).

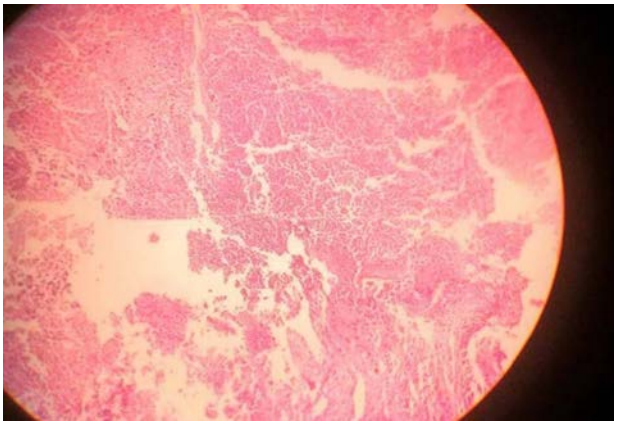

Fig. 3 Trans conjunctival biopsy of recurred orbital mass showing highly undifferentiated squamous cell carcinoma features

In the view of the spread of the tumor, the orbital exenteration was planned under general anesthesia with the ENT surgeon's assistance. Since lids were uninvolved, lid-sparing exenteration was planned. Incisions taken on the orbital margins and the periosteum were elevated all around the orbital walls up to the orbital apex. Stump at the apex was ligated, excised and the bare orbital walls were covered with thigh skin. An orbital conformer prepared from a plaster of Paris was placed in the orbital cavity to improve cosmetics.

Results

An exenterated mass showed features of rhabdomyosarcoma on histopathology (**Fig. 4**). Postoperative period was uneventful and the patient was referred to a higher institution for orbital chemoradiotherapy.

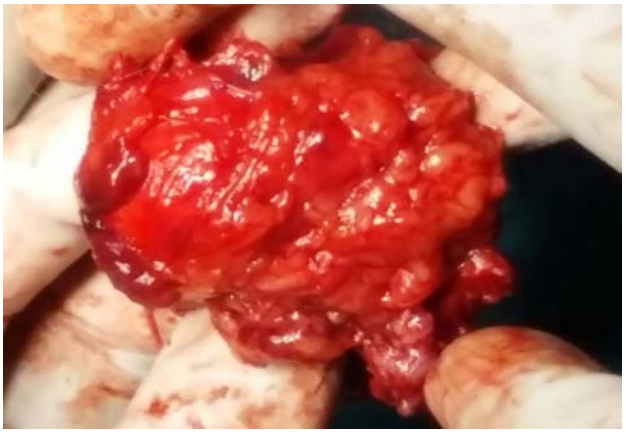

Fig. 4 Exenterated recurred mass from the orbit

Discussion

We presented the case of inverted papilloma of paranasal sinuses, which have been treated eight years before by transnasal functional endoscopic sinus surgery and now presenting with an acute proptosis for 15 days, while the patient was apparently healthy after an initial tumor resection. The patient was subjected to post excisional radiation as the histopathological report revealed features of squamous cell carcinoma. Then, the patient was not followed up for eight years after receiving radiotherapy.

Although sinonasal squamous carcinomas are managed preferably with a comprehensive surgical resection with intra operative frozen section for margin control, in the present case study, a complete tumor resection was performed through an endoscopic approach, as there was no transorbital tumor extension, which could include the removal of some amount of the normal tissue. The meta-analysis on 400

inverted papillomas revealed that the endoscopic surgery was a reliable alternative to traditional external techniques and may be contraindicated in the presence of a transorbital extension in the majority of the lesions [**5**].

The clinical diagnosis of recurrence was considered as the orbital mass was irregular, fixed, and hard in consistency and bled when being touched during the incisional biopsy. An immobile mass was fixed to bone and a painful proptosis with extraocular muscle infiltration provisionally suggested a tumor recurrence. The biopsy material was subjected for histopathology and immunohistochemistry revealing a highly undifferentiated cytological picture featuring squamous neoplasia and a doubtful presence of small cell carcinoma, plausibly carcinoid tumor respectively. An exenteration was performed for fear of rapidity of tumor growth dissemination under general anesthesia and the whole stump was excised. The histopathological examination of the stump showed features of rhabdomyosarcoma.

The recurrence material after the incisional biopsy was sent for immunohistochemistry and revealed a doubtful presence of small cell carcinoma, probably carcinoid at origin, the histopathology showing features of squamous cell carcinoma (**Fig. 5**). Post incisional biopsy suggested squamous cell carcinoma features; however, immunohistochemistry did not report any confirmatory evidence on the cell type, which suggested a probable diagnosis of small cell neoplasm.

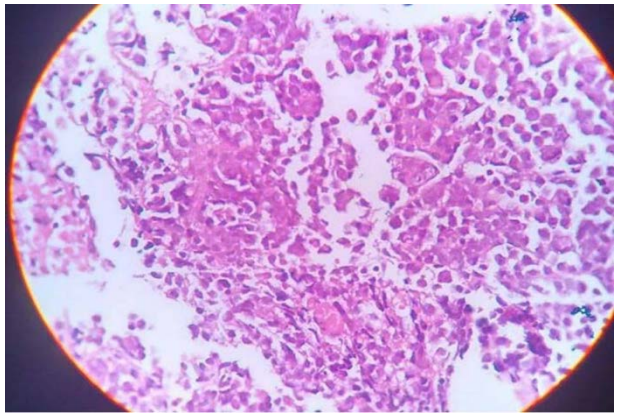

Fig. 5 Exenterated mass revealing features of rhabdomyosarcoma changes on histopathological section

As the exenterated mass on histopathology revealed features of rhabdomyosarcoma that were confirmed, attempts to the subject's recurred mass for immunohistochemistry seemed optional in addition to the financial constraints from patient's side. As the histopathological features were consistent and confirmatory with those of the rhabdomyosarcoma picture, no immunostaining was tried for its verification because histopathology was usually considered confirmatory evidence.

In a retrospective study of 66 patients with IP, treated by endoscopic sinus surgery, 71.5% recurred in the first 12 months compared to the present study of eight years of recurrent free life [**6**]. The structural involvement and the CT findings were correlated with stage II-III Krouse's classification at the initial presentation. After the recurrence and rhabdomyosarcoma features, the disease was categorized under stage IV [**7**].

In contrast to the transitional cell transformation reported by Bajaj et al., the present study revealed a recurred tumor represented as an association rather than a transformation into rhabdomyosarcoma because the histopathology examination did not show the original cell type of the IP [**8**]. From a retrospective study of 23 patients, the recurrence period ranged from 1 month to 14 years after the initial surgery [**9**].

In another retrospective study of 22 patients, the recurrence occurred in 5 cases after an average of 26 months and reported a malignant transformation to squamous cell carcinoma [**10**]. Sung Jin Lee described a patient with pleomorphic rhabdomyosarcoma accompanied with inverted papillomas [**11**].

In conclusion, inverted papilloma after the initial resection manifested as orbital metastasis after eight years to produce an acute proptosis. The recurred material showed features of SCC in an aggressive mode in the biopsy material and on the histopathological section, the exenterated mass revealed rhabdomyosarcoma features subsequently representing as an association or a new primary rather than a transformation as the initial original cell type of IP was not observed in the recurred mass on the cytological examination. Regarding our best literature search, this was the first case report of

rhabdomyosarcoma, presenting as an association with inverted papilloma as evidenced by the histopathological examination.

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Disclosures
None.

Conflict of interest
None.

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