

Case Report

Inverted papilloma presenting as unilateral proptosis: A case report with review of literature



Poonam Sagar^{a,*}; Ishwar Singh^a; Pragya Rajpurohit^a; Shramana Mandal^b

Abstract

Inverted papilloma is an interesting benign tumour arising from lining epithelium of paranasal sinuses which most commonly involves nasal cavity and paranasal sinuses. However, involvement of orbit and intracranial extension without malignant transformation is very rare. We report a case of extensive inverted papilloma of frontal sinus which primarily presented with proptosis, an uncommon presentation. Ophthalmologic symptoms are rare manifestations of paranasal sinus inverted papilloma without malignant transformation and signify extensive disease with possible intracranial extension.

Keywords: Proptosis, Frontal sinus, Inverted papilloma

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Introduction

Inverted papilloma also known as Schneiderian papilloma is a unique tumour characterized by proliferation of epithelium into the stroma of tumour. Inverted papilloma has propensity for local invasion and tendency to recur. Though an intermediate grade tumour, inverted papilloma may undergo malignant transformation.¹ Inverted papilloma usually present with nasal obstruction and discharge. Eye symptoms are very rare and signify extensive disease. We present a case of benign inverted papilloma with intracranial and intraorbital extension, primarily involving frontal sinus which presented as proptosis with appropriate review of literature.

Case report

A thirty two-year-male was referred from ophthalmology & neurosurgery department with chief complaints of right

eye proptosis and swelling for one year. Right eye swelling was insidious in onset and gradually progressive. It was associated with gradual diminution of vision for four months. There was no history of diplopia. Patient also developed right sided partial nasal obstruction for three months which was gradual, continuous and was associated with few episodes of nasal bleeding. There was no history of nasal discharge and hyposmia. On clinical examination, patient had right eye non-axial proptosis with eye ball pushed infero-laterally (Fig. 1). Visual acuity was 6/18 and visual field index was 98%. Pupillary reaction showed right eye relative afferent pupillary defect (RAPD). Nasal endoscopic examination showed fleshy polypoidal mass high in right nasal cavity till the level of middle turbinate with deviation of nasal septum toward left side. Magnetic resonance imaging (MRI) brain and orbit showed altered signal intensity lesion involving right frontal sinus with extra-axial extension into the anterior cranial fossa. The lesion also extended into the right orbit

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^a Department of ENT & Head and Neck Surgery, Maulana Azad Medical College & Associated Lok Nayak Hospital, Delhi, India

^b Department of Pathology, Maulana Azad Medical College & Associated Lok Nayak Hospital, Delhi, India

* Corresponding author at: C-11A, Ram Dutt Enclave, Uttam Nagar, Delhi 110059, India.
e-mail address: poonamsgr.mamc@gmail.com (P. Sagar).



Fig. 1. Clinical image showing globe displacement and eye swelling.

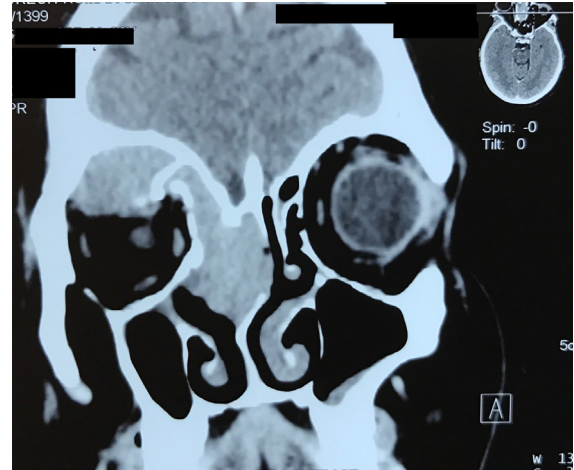


Fig. 2b. CECT brain and PNS (coronal section) showing enhancing lesion involving frontal and ethmoid sinuses with erosion of floor and orbital extension.

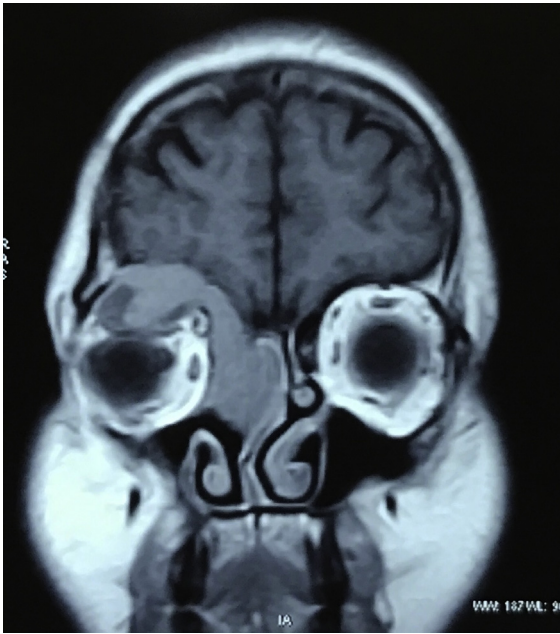


Fig. 2a. CEMRI brain, orbit and PNS (coronal section) showing altered signal intensity enhancing lesion right anterior cranial fossa, orbit, frontal and ethmoid sinuses with deviation of eyeball infero-laterally.

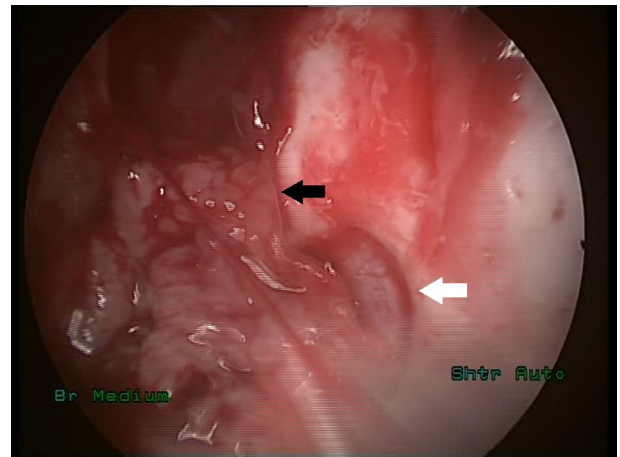


Fig. 3a. Intra-operative picture showing tumour (black arrow) in frontal sinus and defect in the posterior wall of frontal sinus with exposed dura (white arrow).

through medial and superior wall of orbit. There was secondary axial proptosis and mild compression of superior oblique/ superior rectus and levator palpebrae superioris muscle. The lesion also extended into the right ethmoid air cells expanding the right nasal cavity with post contrast enhancement (Fig. 2a). Contrast enhanced computed tomography (CECT) brain and paranasal sinuses showed enhancing expansile mass lesion involving right frontal sinus with orbital and anterior cranial fossa extension (Fig. 2b). Biopsy from the right nasal cavity mass revealed inverted papilloma. Excision of inverted papilloma was done via external fronto-ethmoidectomy and medial maxillectomy via lateral rhinotomy with endoscopic assistance. Intra-operatively tumour was found filling the right frontal sinus breaching inferior wall and extending into the retro-orbital space and around optic nerve. There was dehiscence of posterior wall of frontal sinus with exposed dura (Fig. 3a) but no gross dural invasion, the tumour was carefully elevated off the dura and removed. After gross tumour removal, endoscopic assisted

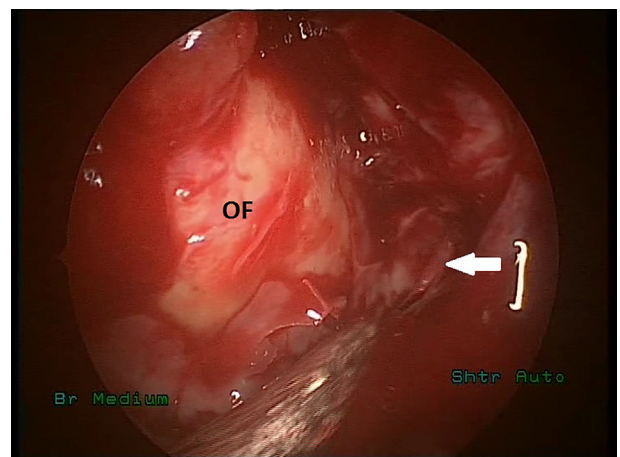


Fig. 3b. Intra-operative picture showing retroconal orbital fat (OF) with dehiscence of lamina, tumour was removed from the posterior part around the optic nerve (white arrow).

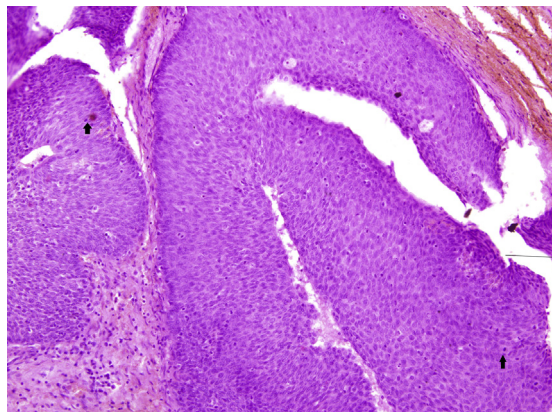


Fig. 4a. H & E section (400X) showing frequent mitosis (black arrows).

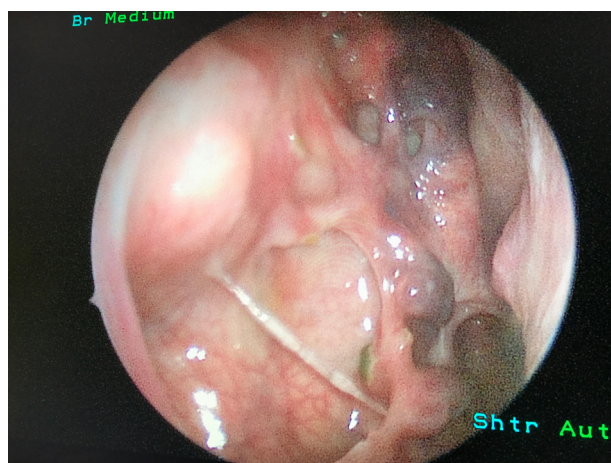


Fig. 4b. Post-operative picture showing well healed nasal cavity and medial orbit.

tumour removal was done from the lateral most part of frontal sinus and around the optic nerve (Fig. 3b). Postoperative period was uneventful. Patient had marked improvement in visual acuity (6/6), proptosis and visual field study. Histopathology showed inverted papilloma with frequent mitosis (Fig. 4a). After one year, patient developed recurrence in frontal sinus which was removed via external approach and patient is being currently under follow-up (Fig. 4b).

Discussion

Inverted papilloma is an intermediate grade tumour arising from Schneiderian membrane of paranasal sinuses. Nasal papilloma was first described by Ward in 1854.² Billroth (1855) described first case of true papilloma of nasal cavity as villiform cancer.³ Hopmann (1883) classified them as hard and soft papillomas.⁴ Ringertz described the true inverting nature of hyperplastic epithelium into the stroma.⁵ Hyams classified papilloma as (1) fungiform (exophytic) papillomas (2) oncocyctic schneiderian (3) inverted papilloma.⁶

Inverted papilloma occurs in 5th-6th decade of life and males are more commonly affected than females. Inverted papillomas constitute 0.4–4.7% of all nasal cavity tumours. Inverted papilloma is associated with upper aero-digestive malignancy (3.4–9.7%), synchronous more than metachronous.¹ In a large study (101 cases), Vrabec reported ocular symptoms in 5 cases only.⁷

The most common clinical presentation of inverted papilloma is unilateral nasal obstruction, discharge and epistaxis. Proptosis, diplopia, diminution of vision, epiphora and headache occur rarely and signifies intra-orbital and intra-cranial extension. Inverted papilloma most commonly arises from lateral nasal wall in the middle meatus region. Ethmoids and maxillary sinuses are most commonly involved, frontal

Table 1. Clinical features, involvement & outcomes of eight reported cases of inverted papilloma with ophthalmic manifestations.

Author	Age/sex	Clinical features	Previous surgery	Involvement	Management	Outcome
Poramate et al. ¹¹	72Y/M	Proptosis, nasal obstruction, discharge & epiphora	Yes	Ethmoids, medial orbit, sphenoid, nose	Excision via LR with eyeball preservation	At 2 years recurrence, death
Wright et al. ⁹	32Y/M	Proptosis, left nasal obstruction	No	Ethmoid, frontal sinus, ACF, orbit with mucocele of maxillary and sphenoid sinus	Endoscopic FE and bifrontal craniotomy	Disease free 1 year
Vural ¹²	51Y/F	Recurrent tumour & ophthalmoplegia	Yes	Intracranial & intradural disease	CFR	Died 2 month post-op
Miller ⁸	42Y/F	Proptosis	Yes	Mucocele frontal sinus. Left frontal sinus, orbit, intracranial with dural involvement	CFR followed by radiotherapy	Eye saved, 3 years disease free.
Peterson et al. ¹³	92Y/F	Proptosis, vision loss, nasal obstruction, discharge, CSF leak, impaired higher cortical functions	No	Nasal cavity, ethmoid, maxillary, frontal pterygoid fossa, MCF, orbit, optic nerve.	No treatment	Died
Van olphen ¹⁴	32Y/F	Swelling left medial eyebrow, b/l nasal obstruction	Yes	b/l nasal cavities, ethmoids, frontal sinus and left ACF	Bifrontal craniotomy, LR & SL approach	Post-op uneventful
Myers et al. ¹⁵	66Y/F	Diplopia, nasal obstruction	No	Medial antral wall, lamina papyracea & frontal sinus	Excision via LR	3 year disease free
Lewis et al. ¹⁶	52 Y/F	Proptosis, nasal obstruction	No	Orbit and intracranial	CFR	6 year disease free

Abbreviations – Y- years, M- male, F- female, LR- lateral rhinotomy, FE- fronto-ethmoidectomy, CFR- cranio-facial resection, ACF- anterior cranial fossa, MCF- middle cranial fossa, b/l- bilateral, SL- sub labial.

and sphenoid sinuses are rarely involved.^{6,7} In the present case, patient had proptosis and eye swelling as primary symptom with primary involvement of frontal and ethmoid sinuses.

Orbital and intra-cranial involvement is very rare in benign inverted papilloma. Our review of English literature revealed eight cases of benign inverted papilloma who presented with eye symptoms (Table 1). Average age was 55.2 years with six females and two males. Our case differs in this aspect, being younger in age. Out of eight cases, four had history of recurrent disease and rest had extensive disease at primary presentation like present case. Various eye manifestations include proptosis, eye swelling, diplopia, diminution of vision and ophthalmoplegia. All the reported cases with eye symptoms in literature were preceded by nasal symptoms except one reported by Miller et al. which also presented with proptosis and mucocele of frontal sinus at primary presentation and on recurrence patient had involvement of frontal sinus, orbit, intracranial and dural involvement.⁸ Our case also had primary involvement of frontal sinus, with tumour eroding inferior wall to extend into intraconal part of orbit and erosion of posterior wall of frontal sinus with intracranial extension into the anterior cranial fossa. Intracranial involvement in inverted papilloma is very rare. Wright et al reviewed eighteen cases of benign inverted papilloma with intracranial extension. Frontal sinus is the most common site for intracranial extension and proptosis is an important sign of intracranial disease.⁹

MRI is an important imaging tool to distinguish between tumour and fluid within mucocele, intracranial and intra orbital evaluation and at present investigation of choice for suspected recurrence. Contrast enhanced computed tomography gives better details of bone invasion and tumour extension.

In present case, tumour was excised via lateral rhinotomy and Lynch Howarth approach removing medial wall of maxilla and external ethmoidectomy. Endoscopic removal of tumour was done from the crevices of frontal sinus, anterior cranial fossa dura, periorbita and optic nerve, preserving its sheath. It is to be emphasized that though inverted papilloma is benign tumour but because of high recurrence rate and local invasion, complete resection is desirable.^{6,7} For intracranial tumours, dural invasion is an important prognostic factor. Lateral rhinotomy, mid-facial degloving, external frontoethmoidectomy, craniofacial resection and endoscopic resection for limited tumours are the surgical options available. Radiotherapy has role in cases of aggressive recurrent disease, intradural invasion, poor surgical candidates and incomplete resection.¹⁰

Inverted papilloma is benign intermediate grade tumour with chances of malignant transformation. Ophthalmologic symptoms indicate advanced disease and possible

intracranial extension. Complete tumour removal is required to prevent recurrence. Intra-orbital extension requires special consideration of preserving eye function. Radiotherapy is reserved for selected cases.

Financial disclosure

None.

Conflicts of interest

The authors declared that there is no conflict of interest.

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