

Case report

Unusual primary sphenoid adenoid cystic carcinoma: the importance of combined CT and MR evaluation

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Objective: To report a diagnostic challenge of primary sphenoid adenoid cystic carcinoma.

Case summary: A 60-year-old premonitory healthy lady presented with progressive deterioration of visual acuity of the left eye associated with diplopia, left epiphora and left ear blockage for two weeks duration. Examination revealed a 'non perception of light' (NPL) of the left eye with left abducent nerve palsy. The nasoendoscopic findings were unremarkable. Initial computed tomography of the brain and paranasal sinus showed a large clivus tumor with intracranial extension while subsequent magnetic resonance detected the epicenter of the tumour appeared to be in the sphenoid sinus with extension to the surrounding structures. A transeptal transsphenoidal biopsy was done reported as mixed pattern adenoid cystic adenocarcinoma. Discussion: Primary sphenoid adenoid cystic carcinoma is an extremely rare slow growing malignancy with non-specific clinical symptoms. The neuro-ophthalmology symptoms are the main presentation. Combined computed tomography and magnetic resonance images are essential in establishing differential diagnosis and to delineate the extent of this disease. Combined modality of surgery and postoperative radiation for adenoid cystic carcinoma has proven better results. **Conclusions:** Primary sphenoid adenoid cystic carcinoma poses a diagnostic challenge clinically. Combined radiological characteristic from computed tomography and magnetic resonance images are essential to aid the diagnosis.

Keywords: sphenoid primary; adenoid cystic carcinoma; salivary gland carcinoma

Bangladesh Journal of Medical Science Vol. 15 No. 02 April'16. Page : 297-302

Introduction

Paranasal sinus carcinoma accounts for about 0.2 to 2 percent of all neoplasm and primary sphenoid malignancy is an extremely rare tumor.^{1,2,4,6} Isolated sphenoid sinus adenoid cystic carcinoma has been always difficult to diagnose because of its location and subtle symptoms. Adenoid cystic carcinoma is an insidious growing, locally invasive tumor of epithelial origin that represents only 1 percent of all head and neck malignancies. The isolated position, difficult accessibility and the tumor characteristic

make great defiance for an early accurate diagnosis. Therefore, high degree of suspicious is needed at the time of presentation; as well as radiological characteristic must be thoroughly reviewed. We report an extremely rare primary sphenoid adenoid cystic carcinoma mimicking clivus tumor with the importance of combined study with both CT and MR images. Highlights on the radiological features, management and the outcome of the case are being discussed.

Case report

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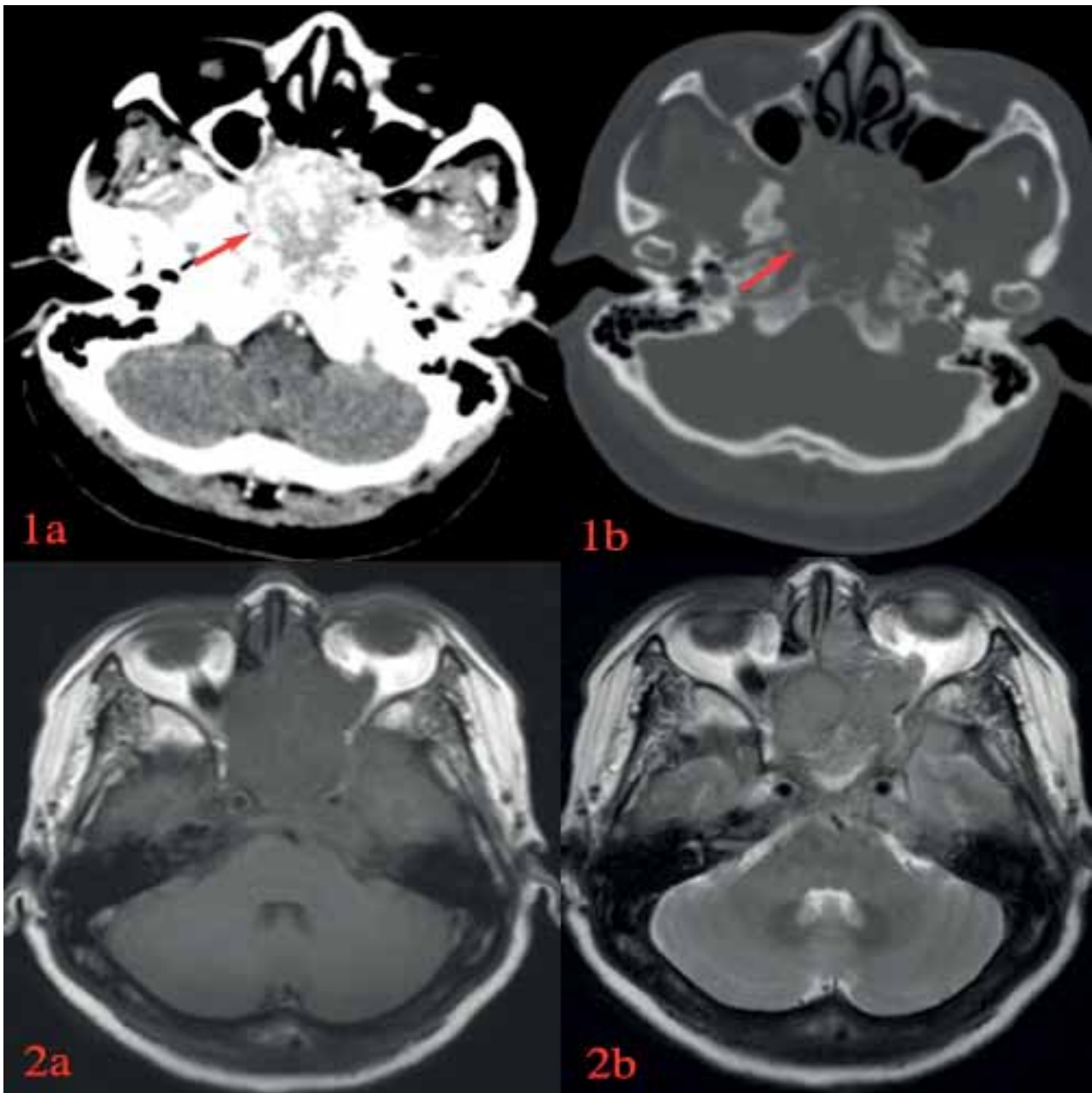


Figure (1) Axial CT images of the brain in soft tissue (1a-arrow) and bone (1b-arrow) settings showing a large enhancing soft tissue mass in the clival region causing erosion and destruction of the surrounding bones while axial MR images of T1 and T2 (2a and 2b) showing the intermediate intensity of the lesion.

A 60-year-old premorbid healthy lady presented with progressive deterioration of visual acuity of the left eye associated with diplopia, left epiphora and left ear blockage for two weeks duration. She also had mild intermitted left hemicranial headache. Otherwise, she denied any nasal symptoms and there was no history of trauma or other illnesses. Examination revealed a 'non perception of light' (NPL) of the left eye with left abducent nerve palsy. The nasoendoscopic findings were unremarkable. As her visual acuity and proptosis persistently worsening, a computed tomography of the brain

and paranasal sinus managed to be done and it showed a large enhancing soft tissue mass within the clivus occupying the sphenoid sinus, the ethmoid air cells and the left orbital apex. There was intracranial extension into the left temporal fossa associated with erosion and destruction of the vomer, clivus, left petrous bone, left medial and lateral pterygoid plates and pituitary floor. Both fossa of Rosenmuller, pituitary and suprasellar region appeared normal(Figure 1). The radiological impression from the scan was a clival bone lesion with differential diagnoses of clival chordoma and

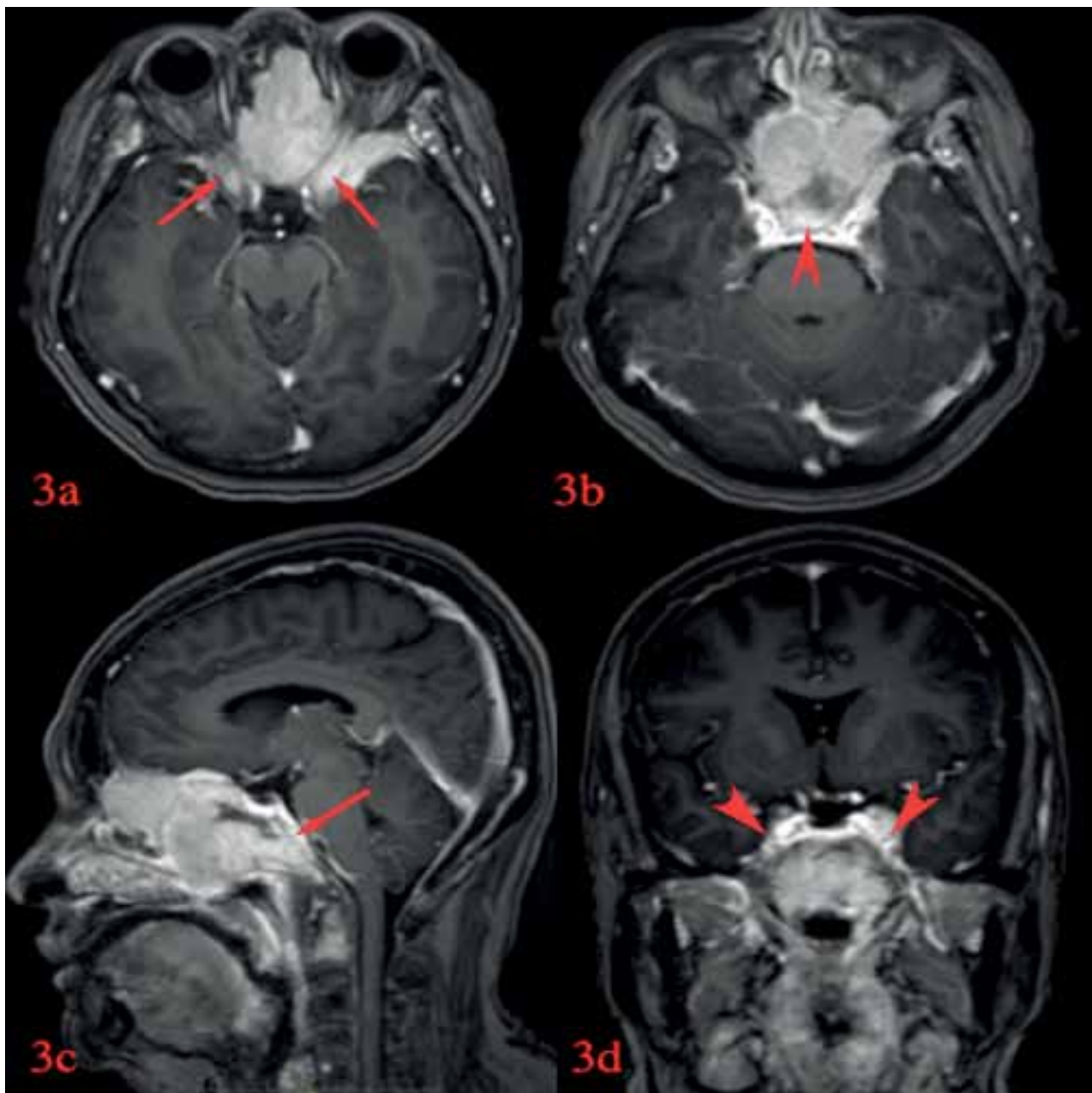


Figure (2) Post gadolinium axial MRI images of the brain showing involvement of bilateral optic nerves and extension the mass into the ethmoidal air cells (3a-arrow and 3b-arrowhead respectively). No extension to the posterior basal cisterns or brainstem demonstrated. Post gadolinium sagittal MRI image (3c-arrow) showing clival involvement but the epicenter noted within the sphenoid sinus and abnormal signal in C2 vertebra. Coronal view (3d-arrowhead) showed bilateral cavernous sinuses involvement.

ethmoid or sphenoid malignancies.

Referral to the Neurology team and magnetic resonance imaging of the brain was arranged. Magnetic resonance images showed a huge midline soft tissue mass occupying and expanding the sphenoid sinus. The mass was isointense to gray matter on both T1 and T2 weighted images, with avid enhancement post gadolinium administration (Figure 2). It extended anteriorly into the ethmoidal air cells and posteriorly to the clivus. There is

obliteration of both optic canals and the mass encased the intracanalicular and intraorbital parts of both optic nerves. Cavernous sinuses on both sides were involved with sparing of the internal carotid arteries. Cranial nerves III, VI, V (ophthalmic and maxillary divisions) and VI were not visualized suggestive of involvement. Abnormal signal intensity of C2 vertebra was also noted possible of bone metastases. The radiological impression was an aggressive lesion arising from the sphenoid

sinus with differential diagnoses of squamous cell carcinoma and adenocarcinoma

Subsequently, a transeptal transphenoidal biopsy was done and histologically reported as adenoid cystic carcinoma with mixed pattern showing arrangement in cribriform and solid clusters. She was planned for surgical resection but she opted to seek alternative medicine. As the disease progress, the symptoms worsened with bilateral complete blindness, ptosis, severe proptosis, swelling of the left eye, left sided headache and the tumor almost encroaching outside the nasal cavity (Figure 3). She was seen at multi disciplinary clinic ORL-oncology and was suggested for palliative chemoradiation as surgical management was not possible.

Discussion

Sphenoid sinus is an extremely unusual site for primary tumor occurrence, with a reported incidence of 0.2 to 2 percent of all paranasal sinus tumors. ^{1, 2, 4, 6} Adenoid cystic carcinoma itself is a rare tumor entity represents only 1 percent of all head and neck malignancies and sinonasal adenoid

cystic carcinoma accounts only 10 to 25 percent of all paranasal malignancies. ^{6, 9} In cases with histologically proven adenoid cystic carcinoma, few histopathologic variants are found. The most frequently found is the cribriform pattern but solid type has more aggressive clinical evolution. ^{6, 9} Other subtypes are mixed (cribriform and solid) and tubular pattern. ⁹

Adenoid cystic carcinoma is characterized by a long clinical course due to its slow insidious growth, but highly invasive with high recurrence rate. ⁷ The tumour has a predilection for early perineural and haematogenous spread, but lymphatic spread to the regional lymph nodes is rare with only 0 to 5 percent. ⁹ Distant metastases at diagnosis is rare with rate of 1 to 3 percent but if occur, most commonly to the lung with incidence ranges from 35 to 50 percent. ^{3, 9} Patients generally present in their fourth and fifth decades with a female preponderance as seen in this case. ^{1, 6, 7}

Lesions in the sphenoid sinus typically result from direct extension of malignant tumours from the



Figure (3) showing deformity of the patient's face with obvious asymmetry, extensive proptosis and hyper-telorism (A). Anterior rhinoscopy showed the tumor almost encroaching outside the left nasal cavity (B) and (C) nearer view of left nasal cavity.

surrounding structures such as the ethmoid sinus, rhinopharynx, clivus, or by growth of invasive pituitary macroadenomas from the sella turcica. Patients with a lesion in the sphenoid sinus often manifests with mild or non specific symptoms until the tumour expands beyond the walls of the sinus.⁷ The non specific symptoms of this tumour could be misleading resulting in wrong diagnosis and delayed management, most frequently treated as chronic sinusitis.⁶

The first presentation of our patient was progressive blurry of vision with diplopia and left VI cranial nerve palsy diagnosed as orbital apex syndrome. Several literatures described ocular problems as initial symptom of primary sphenoid sinus tumour, occurring up to 75 percent of the patients with majority of them having diplopia.^{1,4} This normally result from unilateral abducens nerve involvement owing its long course within the cavernous sinus which is in close proximity to site of tumour. Partial visual loss and ptosis can be explained by tumour extension to cranial nerves II and III.

Patients with primary sphenoid sinus tumours may also present with non specific headache which described as ipsilateral, retro-orbital, or of the vertex similar to our case who complained of left sided headache.^{1,4} This is probably due to the size reduction of the subcribriform sphenothmoidal chamber. The other possibility is due to the involvement of the sphenopalatine nerve or ganglion, or of the middle fossa or cavernous dura. The magnetic resonance and computed tomography images of these lesions play a basic role to define the neoplasm anatomic extension and the adjacent structures integrity. Thus, they are crucial to establish differential diagnosis since the early clinical symptoms are unspecific. Multiplanar reconstructed CT images are sensitive in determining the degree and extent of bony erosion and destruction particularly important in the sphenoid sinus.² These bony changes can be seen in our case, is typical of aggressive lesion in the sphenoid and the absence of bone in this area can expose critical neurovascular risk during surgery. CT images with adenoid cystic carcinoma have also been reported characteristic of hyperdense irregular serrated border that enhances homogenously with contrast.⁷

Magnetic resonance images on the other hand, provides excellent delineation of the tumour local extension by defining the intimate relationship between the tumour and the surrounding structures.

It is superior to other imaging modalities in determining dural invasion, perineural spread, vascular encasement and intracranial spread.

Magnetic resonance images is extremely useful to differentiate tumour from the surrounding inflammations or secretions within the sinuses. Retained secretions or oedema from inflammations demonstrate low T1 and high T2 signal intensities due to high water content. However, variable signal intensities may be observed in chronic cases where certain amount of free water has been observed. Majority of the paranasal sinuses contain highly cellular materials with minimal water content demonstrating low to intermediate signal on both T1 and T2 weighted images.^{2,5} Thus, intravenous gadolinium administration is useful in this instance. Most of these tumours show diffuse intermediate degree of enhancement, whereas the inflamed tissue typically shows avid peripheral enhancement. As for our patient, the main tumour bulk was seen in the vicinity of clivus with poor delineation of local tumour extension on computed tomography images. In the presence of bony erosion and destruction, the features rendered a few differential diagnosis including clival chordoma and sphenoid carcinoma. However, magnetic resonance images provided excellent analysis of tumour in relation to sella turcica, brainstem and the cavernous sinus. The epicenter of the tumour appeared to be in the sphenoid sinus with extension to the surrounding structures. The intermediate T2 signal demonstrated in our case did not correlate with magnetic resonance findings for most chordomas which exhibit very high T2 signal intensity.⁸ The typical 'thumb sign' appearance (posterior projection of the mass indenting the pons) of clival chordomas was absent in our case which showed preservation of the brainstem. In addition, the sparing of the pituitary gland with sellar floor destruction indicated a non-pituitary origin of the tumour.

The adenoid cystic carcinoma treatment consists of four modalities such as surgery, radiotherapy, chemotherapy and combined approach. Single modality has shown poor outcomes while multi-modality treatments with surgery and postoperative radiation has better results.^{8,9} Some recommend tumor debulking followed by adjuvant therapy while others encourage more aggressive surgery in selected cases to achieve maximum local control and improve prognosis.^{3,8,9} The benefit with the use of chemotherapy is not yet defined. Some important prognostic factors for the adenoid cystic

carcinoma are; anatomic location, size, involvement of adjacent structures, cellular atypia grade, surgical margin and the presence of metastasized ganglions. Initial location has significant prognostic value which tumor of the nasal cavity has a better survival than those of the sphenoid.⁹

As this case represents an advanced tumor with extensive local extension, there is no role for surgical excision. Instead patient was referred to oncology team for palliative chemoradiation.

Conclusion

Primary sphenoid adenoid cystic carcinoma is an extremely rare malignancy with non-specific clinical symptoms. The neuro-ophthalmology symptoms are the main presentation in this case due to cavernous sinus involvement. Combined computed tomography and magnetic resonance images are essential in establishing differential diagnosis and to delineate the extent of this disease.

Conflict of Interest: None declared.

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