Tumors affecting primary the sphenoid region - a review of the main lesions

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Authors: R. Haetinger¹, A. V. T. S. R. MONTEIRO², T. A. L. Freddi¹, S. M. B. Mello¹, C. B. A. Ferreira³, D. M. Nunes⁴; ¹Sao Paulo, SP/BR, ²SAO PAULO, SAO PAULO/BR, ³Sao Paulo/BR, ⁴Sao paulo/BR

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Learning objectives

To demonstrate the radiologic characteristics of primary tumors of the sphenoid a very important region of the skull base, due to its location.

Background

The sphenoid is a very critical region of the skull base and a surgical challenge due to its relationship to the optic, trochlear, oculomotor and trigeminal nerves, as well as to the internal carotid arteries, cavernous sinuses, pituitary gland and the basilar artery. Furthermore, from the structural point of view, the sphenoid supports the whole skull and brain (since it is literally the "base"). Many of these tumors are accessible to endonasal endoscopic surgery and the role of the radiologist is to investigate and show all aspects for a safe surgery.

The authors describe and illustrate the main sphenoidal tumors that should be known for the differential diagnosis, based on Computed Tomography and Magnetic Resonance Imaging, which include fibrous displasia, ossifying fibroma, clival chordoma, fibromyxoma and giant cell tumor. Other rare lesions originated in this anatomic site such angiosarcoma are described.

Imaging findings OR Procedure details

Introduction

The sphenoid is a bone at the base of the cranium that is of the utmost importance, since it is the base that sustains the cranium and has a close relationship with the neurovascular structures of the cerebral trunk and the hypophysis, making it a very complex structure and a source of risk for surgical approaches. More than lesions involving other cranial and facial structures, lesions that directly involve the sphenoid bone require an early and precise diagnosis to optimize the chances of their being amenable to treatment.

In the present work, the authors demonstrate the main types of lesions that generally affect the sphenoid bone, and discuss the typical characteristics that they present with during diagnosis by imaging. Lesions coming from adjacent areas that invaded or secondarily deformed the sphenoid were excluded from consideration.
Discussion

Because primary neoplastic lesions coming from the mucosa of the sphenoid sinus are extremely rare, there is no established TNM classification scheme for them. Lesions originating in the sphenoid bone present with a range of diagnoses in pathology, and the various lesion forms cannot always be differentiated from one another solely via radiological methods. Fibrous dysplasias and cement-ossifying fibromas are among the most difficult lesions to differentiate from the point of view of diagnostic imaging. In some cases, their forms are practically indistinguishable in computerized tomography (CT) and magnetic resonance (MR) images. Experience shows that, in pathology analysis, the different lesions are readily differentiable even when viewing only fragments of the lesions. Thus only anatomo-pathological correlation together with radiology images allow for a precise diagnosis. On the other hand, some tumors have highly characteristic imaging profiles that give them a high probability of receiving a proper differential diagnosis. One lesion that causes an osteolytic lesion in the sphenoid, but which has another origin, is the cordoma. Cordomas are of neurological origin and are related to traces of the notochord. Below, the authors describe and give examples of the principal radiological aspects of sphenoid lesions.

FIBROUS DYSPLASIA

Fibrous dysplasias can involve any bone in the body. They occur in the cranium and on the face in a monostotic form in 10 to 25% of affected patients and in a polyostotic form in 50% of affected patients. There are three variants:

- Ground-glass (pagetoid) (56%): characteristically present as substitution of the trabecular of the medulla ossea by tissue mixed with areas of sclerosis and fibrosis, with a gray and relatively homogenous appearance.
- Homogenously dense (sclerotic) pattern (23%): homogenously dense appearance of the medulla ossea.
- Cystic variant (21%): hypodense and round areas surrounded by fine, confluent bone marrow walls.\(^1\)

In CT images, the most common view is of the aspect of expanded bone, with areas of "ground glass", and dense calcifications and/or hypodense areas, with the preservation of the cortical bone. In MR images, a fibrous dysplasia can mimic a tumor. On T1WI, there is a low (more tissue mineralized) or intermediate (fibrous tissue) signal, possibly with high signal areas (collections of medulla ossea). Post-gadolinium T1WI may show enhancement (due to metabolical activity and good vascularization). On T2WI, there is a variable signal with low (more tissue mineralized) and high (non-mineralized areas and cysts) areas.\(^2\)
Fibrous dysplasia lesions are generally painless. If there is pain, other pathologies should generally be considered, unless the lesion is causing compression on the trigeminal nerve such as if it is causing a foraminal stenosis. Another situation that generates symptoms is the rare case of the compression of other cranial nerves such as, in the case of the sphenoid, compression of the optic nerve, which can cause visual changes. Depending on the lesion's location and extent, facial asymmetry can occur.

Fibrous dysplasia is associated with McCune-Albright Syndrome, wherein it generally appears in a polyostotic form (PFD), together with "café au lait" cutaneous maculae and autonomous hyperfunctioning of one or more endocrine glands. According to Matthew et al, the incidence of PFD with this syndrome is 98%. (3) (FIG 1, 2).

**FIBROMYXOMA (MYXOMA)**

Myxomas are rare, accounting for approximately 0.1% of head and neck tumors, with the majority of myxomas affecting the mandible and maxilla in patients with an average age in the range of 25 to 35 years old, with a slight preference for women. They are benign tumors originating in the mesenchyme and generally found in the atria, but can eventually affect bone, muscle, subcutaneous, or skin tissues. When these tumors affect the paranasal sinuses, they can be difficult to diagnose due to nonspecific symptoms, making the diagnosis one of exclusion. They may be restricted to the sinus of origin or may expand to adjacent structures.

Myxomas of the head and neck are rare and, when present, generally affect the mandible and the maxilla. Some cases have been described in which these tumors affect the temporal bone, the pharynx, the larynx, and soft areas. Treatment involves surgical resectioning, with security margins because of the high risk of recurrence.

Myxomas primarily surrounding the maxillary sinuses are described as paranasal sinuses. The first case of sphenoid sinus myxoma reported in the literature involved a 45-year-old male patient complaining of chronic obstruction of the left nostril. Nasofibroscopy showed a whitened lesion with superficial vessels behind the concha media that essentially obstructed the entire left nasal cavity. CT (without contrast medium enhancement) confirmed a mass in the sphenoid sinus that extended across the left ethmoid sinus, the pterygopalatine fossa, and the middle cranial fossa. The lesion promoted erosion of the pterygopalatine layers and remodeling of the posterior wall of the left maxillary sinus, suggesting that it was a slow growing mass. MR showed signs of central necrosis and gadolinium enhancement affecting the middle cranial fossa and the pterygopalatine fossa in the regions of the round foramen and the vidian canal. There were signs of erosion of the floor of the sphenoid sinus and of the posterior wall of the concha media. The biopsy results suggested myxoma, which was confirmed postoperatively. (4)
OSSIFYING FIBROMA

Ossifying fibromas are generally painless benign bone tumors that present with slow growth and are found mainly in the frontal temporal region. This type of lesion is classified as an ossifying fibroma or juvenile ossifying fibroma. The first type is found mainly in women between 30 and 40 years of age. The juvenile ossifying fibroma behaves more aggressively, affecting pediatric patients up to 15 years of age, and has no preference for either sex.

CT imaging features: hypodense circumscribed lesions, with hyperdense points, that can be uni- or multiloculated and have an irregular enhancement by iodated contrast medium. Among the principal differential diagnoses are osteoblastoma, desmoplastic fibroma, and fibrous dysplasia. (5) (FIG 4).

CORDOMA

Cordomas are slow growing, aggressive tumors that originate from remnants of the notochord. They account for less than 1% of intracranial tumors; they are most commonly found in the sacrococcygeal region, followed by the base of the cranium. These tumors are two times more common in men than in women, with a high level of local invasion and high rates of recurrence. In young patients, they occur most commonly in the base of the cranium.

CT usually shows a mass of soft parts with points of calcification that have destroyed the clivus and that can extend to the sphenoid sinus. The lesion presents as an area of hyposignal on T1WI MR and as an area of hypersignal on T2WI MR, with heterogeneous enhancement after endovenous injection of gadolinium. (6) (FIG 5).

GIANT CELL TUMOR

There are two types of giant cell lesions: true giant cell tumors (GCT) and reparative giant cell granulomas (RGCGs). They are indistinguishable by imaging. GCTs are benign neoplasias, with aggressive local potential (in approximately 10% of cases), that primarily affect women, especially in their 40s. They rarely occur in the cranium, but when in this topography, they have a predilection for the sphenoid and temporal bones and can eventually cause disruptive symptoms including headaches and nerve paralysis. (7) The RGCG is a reactive non-neoplastic lesion that is generally related to the history of a progressive trauma, and most often located in the maxillae and mandibles of young
patients. It has a similar histology to the GCT, but with a greater fibrogenic component than that found in true GCTs.

CT shows an expansive lytic bone lesion with well-defined margins, without peripheral sclerosis and with enhancement in the soft tissue component. In MR, giant cell lesions are often isointense to gray matter in the sequences shown on T1WI and T2WI, with intense enhancement by the paramagnetic agent \(^\text{(8)}\) (FIG 6).

**ANGIOSARCOMA**

Angiosarcomas are malignant tumors of endothelial lineage. They are a rare subtype of sarcoma, representing less than 1% of all sarcomas.\(^\text{(9)}\) They occur even less frequently in the cranium, with very few cases having been known to affect the sphenoid bone. They occur in the base of the cranium in patients from 3 months to 50 years of age, with the average age of incidence being around 24 years old.\(^\text{(10)}\)

These tumors have a poor prognosis, with distant metastatic dissemination, resulting in a 2-year survival rate as low as 10% for some subtypes. They are treated by surgical excision; the disease may be best controlled by combining the freezing technique with radiation treatment.\(^\text{(11)}\)

In CT, angiosarcomas appear as osteolytic, infiltrative lesions with irregular and mainly peripheral impregnation by the contrast agent. In MR performed on T2WI, the lesion has a variable signal intensity; and with T1WI post-gadolinium, enhancement is diffuse and heterogeneous. (FIG 7, 8).

**SPHENOID OSTEOMA**

Osteomas are benign bone tumors that are, in the majority of cases, asymptomatic and characterized by slow growth of cortical bone or, less frequently, of cancellous bone. They appear as small and dense lesions generally located in the cranium, more specifically in the frontal and ethmoid sinuses, rarely affecting the sphenoid. In CT, the osteoma lesion has well-defined borders, is homogenously hyperdense, and is generally of small size. In MR, the lesion image is hypointense in all sequences.\(^\text{(12, 13)}\) (FIG 9).

**ANEURYSMAL BONE CYST**

The aneurysmal bone cyst is a non-neoplastic multicystic tumor made up of connected spaces filled with blood. It occurs mainly in young people between 10 and 20 years of age
and is generally located in the long bones and the vertebrae. Only 1% of these lesions occur in the cranium, and they are even more rare in the sphenoid.

In CT, the aneurysmal bone cyst is characterized by a heterogeneous mass, with multiple liquid-liquid levels. In MR, it often appears as a heterogeneous signal in sequences performed on T1WI and T2WI, with septations and a hyperintense capsule as well as liquid-liquid levels. Although we do not have an example of this lesion to present here, we consider it important to mention in the interest of being as complete as possible with respect to the differential diagnosis of sphenoid lesions.

Images for this section:
Fig. 1: A, B, C. FIBROUS DISPLASIA: CT with the coronal bone window on the axial plane (A) showing the sphenoid bone with an expanded aspect and with "ground glass" involving the body of the sphenoid, without the development of left sphenoid sinus and with hypoplasia of the right sphenoid sinus. The coronal plane (B) also shows an expanded aspect in the anterior clinoid processes and the relation of the lesion with the left optical canal (arrow). In the oblique sagittal plane (C), it can be seen that the optical canal is not narrowed, despite the extent of the bone lesion (arrow).

Fig. 2: A, B, C, D: FIBROUS DISPLASIA WITH PNEUMATOCELE. MR images showing T2WI axial (A), T1WI sagittal (B), and T1WI axial (C) views. The signal intensity is predominantly low, as much on T1WI as on T2WI. This case also shows a pneumatocele in the sphenoid coming from the left mastoid, as shown on CT (D).
Fig. 3: A, B. FIBROMYXOMA: CT in the coronal plane with bone window (A) and post contrast media with soft tissue window (B) showing a process of expansion involving the diploe of the sphenoid body and the pterygoid process on the right. Note the irregular central calcifications and thinning accompanied by minor destruction of the cortical bone on the interior side of the sphenoid (arrow). The soft tissue component of the lesion was impregnated irregularly by the contrast medium.
**Fig. 4:** A, B, C, D. OSSIFYING FIBROMA: CT with a soft tissue window of the axial plane (A) showing a process of expansion involving the sphenoid and the maxilla on the right, presenting with hypodense contents permeated with coarse calcifications and with irregular impregnation by the contrast medium, obliterating the adjacent pterygopalatine fossa and invading the masticator space. The coronal plane with a soft tissue window (B) showing the lesion to be contiguous with the right optical canal (arrow) and confirming invasion of the pterygopalatine fossa. An image of the most anterior coronal plane with a bone window (C) shows bulging of the orbital floor and expansion to the ipsilateral ethmoid. In the most central part of the lesion, the bone has heterogeneous areas with a "ground glass" pattern interlaced with hypodense areas of probable fibrous tissue. The cortical bone has been remodeled and has periosteal narrowing in the walls of the paranasal sinuses, while the orbital floor has lost its definition. Fragments of this tumor following removal from the maxilla are shown in Fig.3-D. (Courtesy of Prof. Dr. Lidio Granato (Hospital Santa Casa de Misericordia Sao Paulo, SP, Brazil).
Fig. 5: A, B, C, D CORDOMA OF THE CLIVUS: (A) MR performed on T1WI in the axial plane after gadolinium, showing the process of expansion on the posterior side of the sphenoid, mainly on the left, with heterogeneous impregnation. (B) MR performed on T2WI in the sagittal plane shows the hyperintensive characteristic of this type of lesion, with slightly heterogeneous parenchyma. (C) CT with a bone window in the sagittal plane showing the osteolytic lesion in the clivus, involving the dorsum sellae. (D) A 3D reconstruction with volume rendering giving a good indication of the lesion's topography and of the areas where the clivus has been destroyed.
Fig. 6: A, B, C and D: GIANT CELL TUMOR (GCT): Expansile lesion occupying the topography of the sphenoid body and extending itself backward in the direction of the pre-pontine cistern, laterally dislocating the cavernous sinuses, and forward in the direction of the sphenoid sinuses, compressing them. The tumor restricts diffusion (A), with heterogeneous signal intensity on T1WI pre-gadolinium images and has predominantly peripheral enhancement in the post-gadolinium sequence (C and D).
Fig. 7: FIG 7 A, B, C, D, E: ANGIOSARCOMA OF THE SPHENOID (Part 1): The first CT exam (pre-operative) with axial images with a bone window (A) and the soft tissue post intravenous contrast (B) showing a lesion in the clivus with anterior extension and invading the sphenoid sinuses, but preserving a good portion of the intersinus septum, with intense and irregular impregnation in the most posterior portion. MR performed on T2WI (C) shows tissue with mixed signal intensity occupying the clivus and both of the sphenoid sinuses (preserving the intersinus septum). MR performed on T1WI post-gadolinium on the axial plane (D) with diffuse and irregular impregnation and on the sagittal plane (E) and with expansion across the whole body of the sphenoid.
**Fig. 8:** FIG 8 A, B: ANGIOSARCOMA OF THE SPHENOID (Part 2): CT performed 2 weeks later (A and B) revealed destruction of most of the sphenoid body; MR confirmed rapid tumor growth, with the same signal characteristics as observed in the previous examination, but larger (MR images not shown here).
Fig. 9: FIG 9 A, B, C: OSTEOMA: CT in the axial (A) and sagittal (B) planes and tridimensional image ("volume rendering") (C) showing a small exophytic bone lesion, which is homogenously hyperdense, located in the right sinus.
Conclusion

The recognition of the primary tumors of the sphenoid bone has significant importance to radiologists and helps to minimize the diagnostic time and to achieve a better management of these lesions.

Personal Information

References


4- Moore BA, Wine T, Burkey BB, Amedee RG, Butcher II RB. Sphenoid Sinus Myxoma: Case Report and Literature Review. The Ochsner Journal 8:166-171, 2008


