Chordoma Arising from Cranial Base Extending to Oropharynx: An Unusual Presentation

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Abstract Chordoma is a relatively rare tumor of the skull base thought to originate from embryonic remnants of the notochord. Chordomas arising from the skull base/clivus are typically locally aggressive with lytic bone destruction. When chordomas occur in an extraosseous location, they may mimic other lesions of the nasopharynx and oropharynx. We present a case of primarily extraosseous chordoma involving the oropharynx in an effort to improve the preoperative diagnosis of this rare tumor among the dental fraternity. In addition, we review regional notochordal embryology to explain this variant tumor location.

Keywords Chordoma · Extraosseous · Skull base/clivus

Introduction

Chordoma is the name applied to tumors derived from remnants of the notochord is a relatively rare tumor of the skull base and sacrum. The most common location for chordoma is in the sacrum; the skull base/clivus region is the second most common location along the course of the notochord [1]. Rarely, skull base chordomas may occur in an extraosseous location. When this occurs,

chordomas may mimic other lesions of the oropharynx and nasopharynx.

These tumors are benign, but extensive local invasion and occasional metastatic spread makes them behave like malignant tumors [2]. Biopsy and histology make the diagnosis. Usually treatment consists in surgery and radiotherapy but more recently proton beam therapy is used and seems to yield better results.

Case Report

A 73-year-old woman sought treatment for a slowly enlarging pharyngeal mass that had been present for at least 2 months; the pain started 2 months before, gradually increased day by day (Fig. 1a). During this time, she complained of hypernasal speech, dysphagia. Her past history revealed a case of right squamous cell carcinoma (post treatment) right hemimandibulectomy done 10 years ago. The patient reported that she recently had noticed shortness of breath on protruding her tongue. Otherwise, she was normal. On physical examination, the patient appeared moderately built and nourished. The nasal and nasopharyngeal examination revealed no abnormalities. Intraorally, there was an erythematous area in the left soft palate, extending up to midline on same side. Swelling was firm, tender, and sub mucosal mass filling the pharynx (Fig. 1b). There was cervical lymphadenopathy. Keeping in mind post treatment squamous cell carcinoma; we came to preliminary diagnosis of any secondaries, salivary gland tumor, metastatic lymphnode, parapharyngeal carcinoma. Patient was subjected to few preliminary investigations, which included PA maxilla, and water's view (Fig. 2) showed an evidence of large soft tissue swelling in the left side of the soft palate region and pillars of left tonsillar

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Fig. 1 a Clinical photograph of patient. b Intraoral photograph

Fig. 2 Waters view showing haziness of the left maxillary sinus region

fossa. The mass was well defined, smooth in contour, with no evidence of calcification. Left maxillary antrum was dense. OPG showed evidence of right hemimandibulectomy (Fig. 3).

A base skull and neck CT scan revealed a prominent hypo dense globular soft tissue mass lesion noted in the left parapharyngeal area, distorting the adjacent air space, and eroding the pterygoid lamina and displacing the pterygoid muscle, no calcifications, with extension into infra temporal fossa suggesting chordoma (Fig. 4a, b) and accessory parotid tumor. Gelatinous material aspirated from the swelling. The cellular histologic finding from a fine-needle aspirate of this lesion showed myxoid material with

Fig. 3 Orthopantomograph showing right hemimandibulectomy and radiopacity of the left maxillary sinus region

Fig. 4 a Coronal CT showing globular soft tissue mass in the left parapharyngeal area. b Axial plane CT showing globular soft tissue mass in the left parapharyngeal area

interspersed cells, having abundant bubbly cytoplasm. Nuclei are small, and round, occasional one showing conspicuous nucleolus (Fig. 5). Trucut biopsy with 16 gauge needle was performed to confirm the diagnosis (Fig. 6) which was consistent with a chordoma. The patient declined treatment because of her age, financial constraints and the potential adverse effects of surgery and radiotherapy.

Discussion

Chordoma was first described by Luschka in Virchow's laboratory in 1857 [3]. The name was given due to microscopic appearance of the tumor. Chordomas are malignant neoplasm's representing 1% of all malignant bone tumors and intracranial tumors and 0.2% of all naso

and oropharyngeal masses [4]. Embryologically, this tumor derives from vestigial notochord tissue. The theory of the notochordal origin of this tumor was generally accepted following the valuable work stated by Rippert in 1894 [5]. The understanding of these lesions is aided by the knowledge of the embryology of the primitive notochord, with the help of diagrammatic representation of the notochordal remnants (Fig. 7). By the third week of gestation, the notochord begins to develop. The notochord's cranial limit is Rathke's pouch, and therefore it lies at the level of the sphenoid. Inferiorly, it comes in contact with the pharynx and becomes enveloped by the developing vertebral bodies along the entire length of the spine. By the eighth week of development, the notochord involutes, although it does form the nucleus pulposus. Incomplete regression of

Fig. 5 H and E stain showing myxoid material with interspersed cells, having abundant bubbly cytoplasm with vacuolation, occasional one showing conspicuous nucleolus characteristic of chordoma

Fig. 6 Section of trucut biopsy showing physaliferous cells vacuolated, bubbly cytoplasm characteristic of chordoma 40X

Fig. 7 Diagrammatic representation of notochordal remnants

notochordal tissue can result in a chordoma. The notochord persists in the intervertebral discs where it swells up and forms part of the central core. The notochordal element escapes envelopment by cartilage and bone in three places at the cranial end: [1] Between the cartilage and perichondrium of the dorsum sellae; [2] above the occiput towards the Odontoid process; [3] between the base of the skull and pharynx. The clinical picture of a chordoma depends on whether the growth extends upwards into the skull or downwards into the nasopharynx [6].

Chordomas may occur at any site along the course of the embryonic notochord. Most commonly found in the sacrococcygeal region (50%), followed by the skull base (35%), especially in the clivus and the vertebrae (15%) [1]. Intracranial chordomas most often originate from the sphenoid-occipital synchondrosis of the clivus. The site of origin may be along the upper clivus (basisphenoid) or along the caudal margin of the clivus (basiocciput) (Fig. 6). Other sites of origin include the sellar area, sphenoid sinus, and, rarely, the nasopharynx, maxilla, paranasal sinuses, or intradural region [7].

Chordomas affect male patients at a greater rate than female patients. They can occur at any age, the greatest frequency of chordomas in the head and neck region appear during the fourth decade [8]. We have reported a case of female patient aged 70 years, so we can consider it as a rare case.

The clinical signs and symptoms are variable with involvement of base/clivus and one or more of the following may be present: headache, diplopia, paresis of facial and tongue muscles on side of involvement defects of speech and difficulty in swallowing, a troublesome stiff neck and some loss of balance. The signs include the presence of a central pharyngeal mass projecting into the vault of the nasopharynx where it grows to produce nasal obstruction and deafness, paresis of the various muscles of the eyeball, face and tongue, impairment of speech and dysphagia. In the rare oropharyngeal presentation, there will be dysphagia and an altered voice as this was consistent with our patient [9]. X-ray films may reveal a lobulated soft tissue mass projecting into the vault of the nasopharynx as in our patient. The classic appearance of extraosseous chordoma is well-circumscribed, lobular hypoattenuated, expansile soft-tissue mass that arises from the clivus with associated extensive lytic bone destruction, sometimes dystrophic calcifications [10]. These classic features were present in above case.

Intracranial chordomas generally occur often in the region of the sphenoid-occipital synchondrosis. There have been few extraosseous chordomas reported in the literature, but only very few have been reported in the oropharynx, as defined in our case report [8, 9].

Diagnosis of a chordoma could be a challenging task, because of the transient and vague nature of signs and

symptoms of these lesions. The classic differential diagnosis includes chondrosarcoma, clival meningiomas, plasmacytoma, lymphoma, craniopharyngeomas, and rhabdomyosarcoma. Other differential diagnoses, although rare, include aggressive pituitary adenoma, histiocytosis X, dermoid and epidermoid cysts, trigeminal neuroma, and fibrous dysplasia. The differential diagnosis list is significantly different for extraosseous chordoma from the standard classic chordoma; due to its location. The differential for this variant lesion includes nasopharyngeal carcinoma and non-Hodgkin lymphoma [11].

Definitive diagnosis of these well-defined, lobulated, extraosseous tumors requires histopathologic analysis. Chordomas have been divided into two histopathologic subtypes: typical chordomas and chondroid chordomas. In typical chordomas, the cells tend to be arranged in cords set in a pale matrix of mucopolysaccharide with a characteristic physaliphorous appearance. In chondroid chordomas, the stroma resembles hyaline cartilage with neoplastic cells in lacunae. This variant is more commonly seen in the skull base [12]. The characteristic cytoplasmic and intercellular vacuolation is prominent and vacuolated cells may assume signet-ring forms, while in others multiple vacuoles produce a frothy appearance of physaliferous cells [13]. These vacuolated cells are the result of continual aging process. Our patient's needle biopsy and Trucut biopsy showed myxoid material with interspersed cells, having abundant bubbly cytoplasm, which is classic picture of chordoma and with the help of CT we arrived at the diagnosis. With the help of immunohistochemical studies, these tumors can be identified on the basis of elements (epithelial markers) that indicate their relation to notochordal mesenchyme, which are positive for cytokeratin and S-100 [14].

As the lesion was extensive and in close proximity to the brain (base skull), total excision was ruled out. Combined treatment modalities include, surgical removal, chemotherapy, and fractionated proton beam radiation therapy [15]. Local recurrence is still common regardless of the mode of therapy, but distant metastasis is rare. MR imaging is the modality of choice for post surgical follow-up and detection of recurrence [16].

Intracranial chordomas are rare midline tumors of clival origin. Rarely, skull base chordomas may occur in an extraosseous location. When this occurs, chordomas may mimic other lesions of the oropharynx and nasopharynx. These extraosseous lesions are atypical relative to the usual presentation of clival chordoma due to their oropharyngeal location. It is important to consider extraosseous chordoma in the differential diagnosis of tumors in the nasopharynx and oropharynx because it requires a very different treatment plan and it has its own prognosis. Recent treatment modality is based on proton emission, which has proved to be efficient with fewer side effects.

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