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Case Report

Chondrosarcoma of the Nasal Septum: A Case Report and Literature Review

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Abstract

Chondrosarcoma, a malignant neoplasm, is characterized by the formation of a cartilaginous matrix by the neoplastic cells and is associated with a high recurrence rate. Chondrosarcoma of the head and neck are rare, accounting for less than 12% of all cases of chondrosarcoma. Head and neck chondrosarcoma usually involves the maxilla and rarely affects the nasal septum, with only 50 cases reported in the literature. Most head and neck chondrosarcoma occur in people in their forties, with a slight predilection for male patients. Patients usually present with a painless swelling or nasal obstruction. Wide surgical resection is the preferred treatment for chondrosarcoma; radiotherapy and chemotherapy are usually alternative options. Few clinical series have evaluated chondrosarcoma's biological behavior and surgical outcomes involving the head and neck. Here, we report the clinicopathological characteristics of a massive nasal septal chondrosarcoma, probably the largest ever reported in a young girl, resulting in bilateral visual loss and eroding the nasal and oral cavities necessitating a tracheostomy and feeding jejunostomy. Her late presentation, combined with the unusual aggressiveness of the tumor, precluded any attempts at surgical resection.

Keywords: Chondrosarcoma; Head and Neck; Nasal Septum; Surgery

Introduction

Chondrosarcoma is classified by the World Health Organization as a malignant tumor characterized by cartilage-forming, but not bone-forming, tumor cells [1]. The chondrosarcoma in the head and neck is a rare entity, with an incidence of approximately 5% to 10% [2,3]. Chondrosarcoma tends to progressive local destructive extension with frequent recurrences and is considered to have a poor Prognosis [4]. This slow-growing tumor originates from cartilaginous tissues. We experienced in our center an unusually massive, probably the largest ever reported, primary chondrosarcoma of the nasal septum, which eroded the orbits, sinonasal and oral cavities with extension to the anterior cranial fossa and cavernous sinus, causing loss of vision in both eyes. Primary chondrosarcoma of the nasal septum region rarely erodes and disfigures the facial structure or extends to the cranial or intracranial areas unless there is tumor recurrence [5]. Treatment is primarily surgical; adjuvant therapy is recommended if a radical resection is not possible [6].

Case Report

A 17-year-old girl from a small village presented with a vast, irregular, and fungating nasal mass that measured approximately 11 × 8 × 5 cm and disfigured her entire face (Figure 1A, 2A). The mass resulted in blindness in both her eyes. She first experienced symptoms due to the progressively growing mass about 18 months before visiting our maxillofacial surgery clinic. Initially, she had complaints of nasal blockage and frontal headache. Seven months prior to her presentation, she started to have a painless facial swelling with dysphagia and speech difficulty episodes. One month later, she lost vision in her right eye and began to experience blurring of vision in the left eye. The mass had grown aggressively during the last two months before her presentation to our department and started to destroy her facial features. She began to have difficulty breathing, severe dysphagia, inability to maintain oral intake, and speaking difficulties. A history of ear pain was also noted. No detailed information regarding seeking other medical advice was available. Her face appeared completely disfigured with the considerable mass flattening her nose, covering and eroding her eyes, invading her oral cavity, and deviating and destroying her mouth. A large fungating ulcerated lump had eroded through her right cheek. Fundoscopic examination of her eyes showed optic disc atrophy.

A computed tomographic (CT) scan was ordered and performed using a Siemens Somatom Definition AS+ scanner with intravenous contrast (IV) administration. Post-processing and volume rendering was performed using commercially available software (TeraRecon, iNuition Viewer). The scan (Figure 3) showed a massive lobulated soft tissue lesion with multiple islands of chondroid matrix against a minimally enhancing background of soft tissue density. The mass was centered about the nasal septum causing marked widening of the nasal cavity and loss of all anatomical landmarks, including the ethmoid sinuses. Posteriorly, the mass was shown to infiltrate the sphenoid sinus with the destruction of the pterygoid plate. Laterally, the mass was seen to involve the medial orbital margins as well as the orbital apices, resulting in significant compression of the optic nerves at the apex and divergent displacement of the orbital contents and with atrophy of the globe, which was more severe on the right (Figures 3 and 4). Inferiorly, the mass infiltrated the maxilla and hard and soft palate and could be seen as a fungating mass protruding into the oral cavity (Figure 5). Superiorly, the mass was noted to extend to the cribriform plate of the ethmoid with minimal intracranial extension (Figure 6). Anteriorly, the mass was shown to extend beyond the facial bones' margins in an exophytic manner (Figures 7).

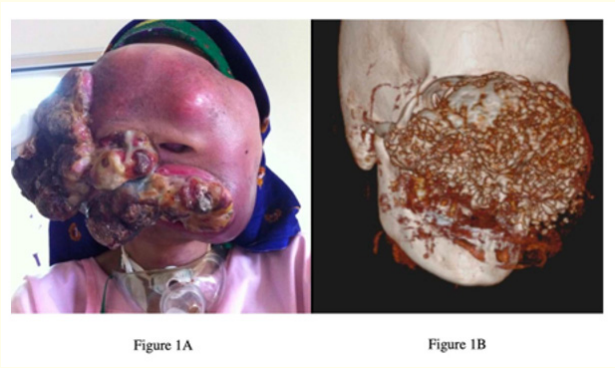


Figure 1A and 1B: Frontal profile view of the patient's face (1A) showing massive distortion of facial features with corresponding 3D Volume rendered (1B) images showing the mass with significant destruction of the facial anatomy.

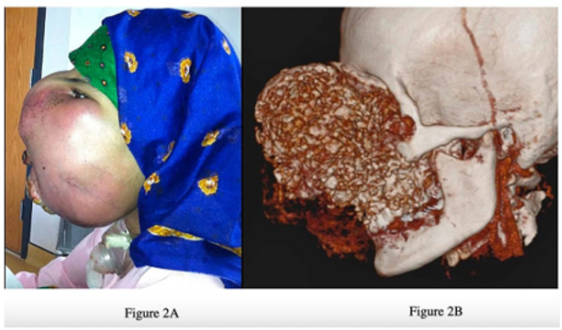
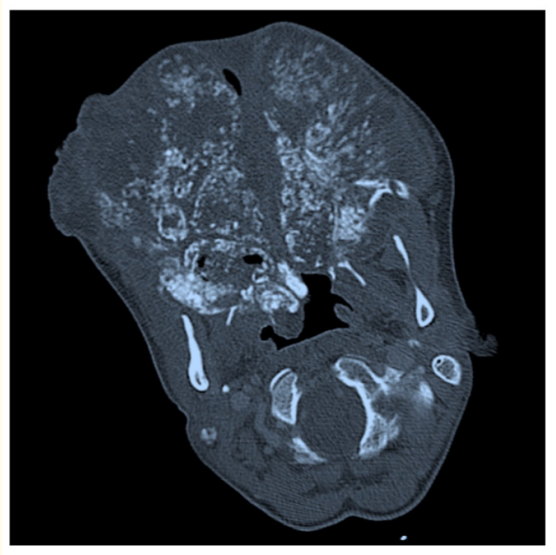


Figure 2A and 2B: Lateral profile view of the face showing the large mass with deviation of the eyes and complete distortion of the normal features of the face with corresponding 3D volume rendered images showing the mass invading into the nasal septum, orbits and mandible.

The volume-rendered 3D images (1B and 2B) demonstrated the deformity of the facial features of the patient (1A and 1B). The



Figures 3: Axial contrast-enhanced axial CT scan of the face in bone window showing a massive, lobulated soft tissue lesion with multiple islands of chondroid calcifications against a background of low-attenuating minimally enhancing soft tissue matrix.

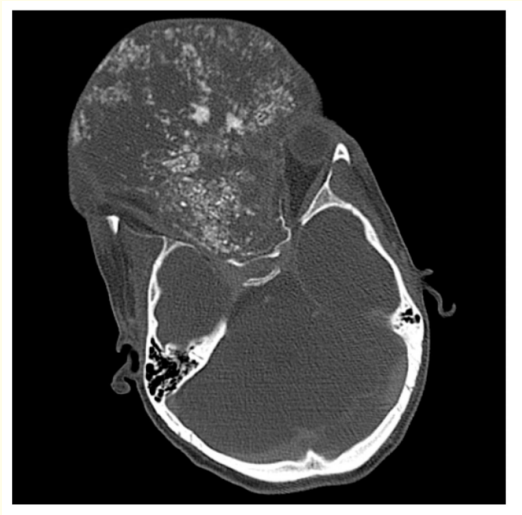


Figure 4: Axial contrast-enhanced axial CT scan, at the level of the nasal septum showing complete destruction of the nasal septal anatomy, bilateral orbital extension and exophytic extension beyond facial boundaries.

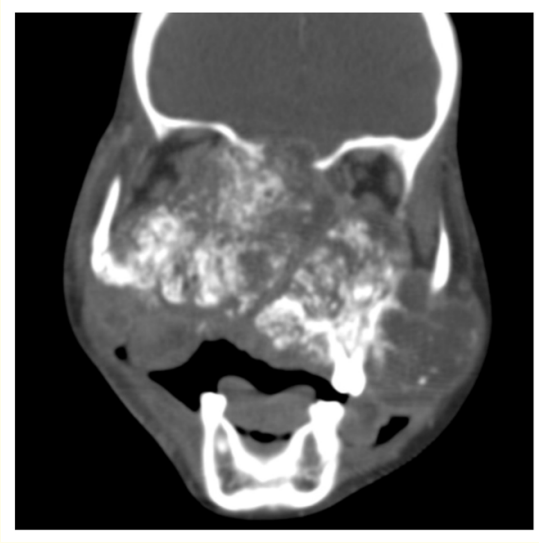


Figure 5: Coronal contrast-enhanced CT scans through the orbits. The mass extends laterally and posteriorly, infiltrating the medial orbital margins and the orbital apices. This has resulted in marked compression of the optic nerves at the apex and divergent displacement of the orbital contents, with globe atrophy, which is more severe on the right.

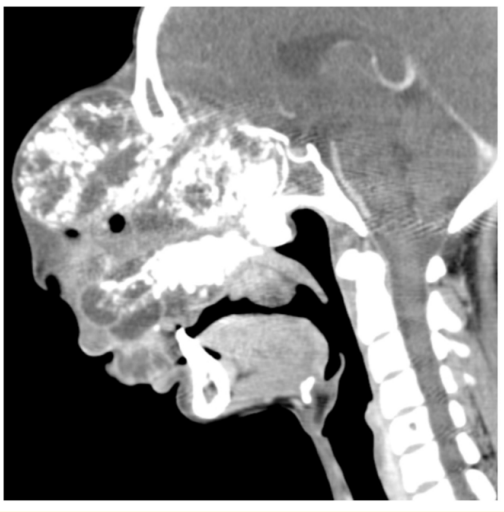


Figure 6: Sagittal contrast-enhanced CT scan shows that the mass has infiltrated the maxilla, and hard and soft palate, and extended into the oral cavity as a fungating mass.

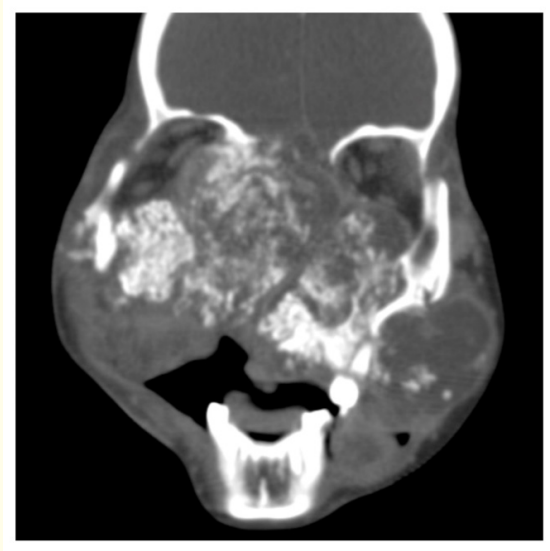


Figure 7: Coronal contrast-enhanced CT of the face shows infiltration across the cribriform plate of the ethmoid with some intracranial extension.

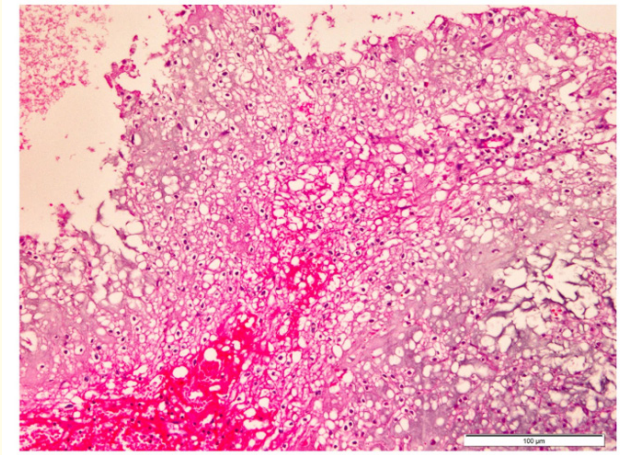


Figure 8 : Representative image of a histopathology slide showing Hematoxilin and Eosin (H&E) stained cells at 4x magnification The slide shows hypercellularity of small round cells and islands of well-differentiated hyaline cartilage.

patient underwent an incisional biopsy, and a histopathological examination revealed moderately to a densely cellular proliferation of a cartilaginous nature made up of multiple confluent lobules. It presents ulcero-necrotic changes, sometimes significant with deposits of fibrin. Tumor cells are small, reasonably monomorphic, with an atypical irregular nucleus, increased in size, and clear chromatin. Bi and multi-nucleations are numerous. Absence of mitosis. In immunohistochemical study, presence of heterogeneous staining of tumor cells with P53. The proliferation index (Ki67) is estimated at 5-10% in hotspot areas. These findings were consistent with a diagnosis of aggressive chondrosarcoma.

The decision to operate was controversial, and a multidisciplinary team (MDT) involving the Otolaryngology-Head and Neck Surgery and Plastic Surgery and Maxillofacial Surgery departments decided that palliative treatment should be administered. A tracheostomy tube was inserted to facilitate breathing, and a feeding jejunostomy tube was inserted for nutritional support.

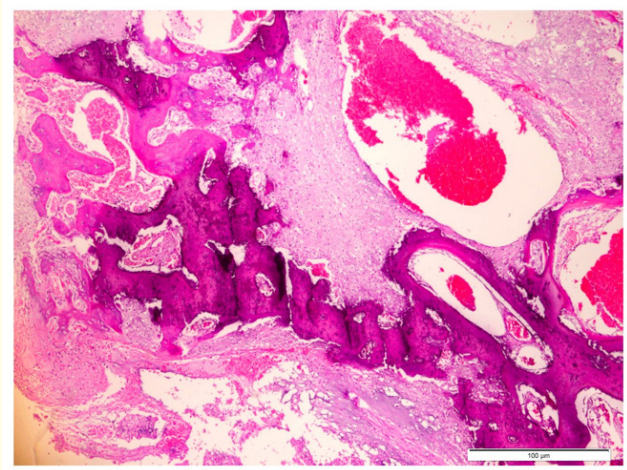


Figure 9: Representative image of a Hematoxilin and Eosin (H&E) stained histopathology slide showing Chondrosarcoma with bone permeation.

Discussion

Chondrosarcoma accounts for 10–20% of all primary malignant bone tumors and, after excluding multiple myeloma, represents the

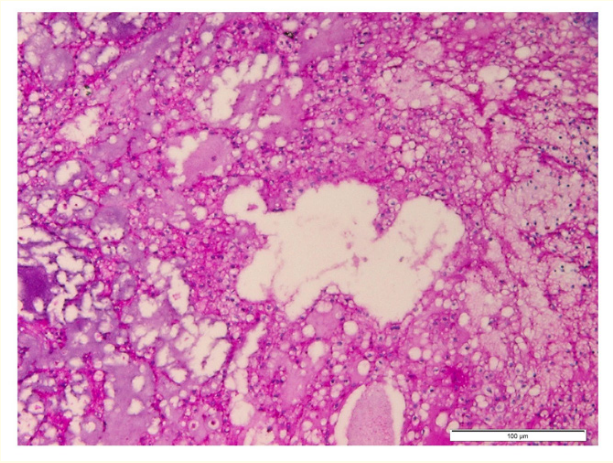


Figure 10: Representative image of a Hematoxilin and Eosin (H&E) stained slide at 10x magnification demonstrating marked hypercellularity in a hyaline matrix.

second most common primary bone malignancy after osteosarcoma [6,7]. Chondrosarcomas rarely involve the head and neck region, accounting for less than 12% of all cases of chondrosarcoma and only 0.1% of all malignant head and neck tumors [8,9]. The most commonly affected sites in the head and neck are the anterior portion of the maxilla, the sinonasal region, and the mandible [10-13]. Moreover, chondrosarcoma represents only 4% of non-epithelial tumors of the nasal cavity, paranasal sinuses, and nasopharynx14. Although this malignant tumor is known to be slow growing, in our patient, it grew to a massive size in a relatively short period, leading to facial destruction and functional abnormalities, which are not typical of chondrosarcoma [1]. Chondrosarcoma may present with a wide range of clinical alterations, but a painless swelling is the most common complaint [15]. In our patient, the first complaint was nasal obstruction and frontal headache, which are the typical symptoms described in the literature. Unfortunately, our patient neglected these early symptoms, probably because of low socioeconomic status, unavailability of good health care, and low educational level. Her late presentation and the aggressiveness of the tumor led her to present with what is probably the largest and most aggressive CHS reported in the literature. Besides its massive size, the tumor was associated with pressure symptoms that have never been reported in the literature, e.g., dysphagia, difficulty breathing, and bilateral loss of vision [5].

Chondrosarcoma is histologically categorized into three grades based on the degree of cellularity, nuclear size and atypia, and mitotic activity. The more aggressive types have highly undifferentiated small round cells and islands of well-differentiated hyaline cartilage [2]. Imaging (MRI and CT) is essential for the characterization of the mass and determining the extent of tumor invasion, which significantly facilitates surgical planning. Typically, CHSs appear as multiple areas of calcification against a background of hypoattenuating matrix on CT scans. The calcifications may be amorphous, large, or occasionally punctuate [3,16]. On magnetic resonance imaging (MRI), the mass appears iso- to hypointense on T1-weighted images and hyperintense on T2-weighted images [16]. Our case is unique in its imaging findings due to the tumor's sheer size and the patient's late presentation.

This neoplasm's most effective treatment modality is aggressive surgical removal with wide resection with negative margins. Since the residual disease is an important cause of recurrence, adequate histologically clear margins must be obtained [9,10,17]. However, in our patient, surgical management was challenging because of the pervasive spread of the tumor.

Furthermore, other available treatment options, e.g., irradiation and chemotherapy, were not applicable because studies have shown that these modalities do not appear to have a significant effect on survival [9,18]. The 5-year survival rate for head and neck chondrosarcoma varies between 32% and 87.5%. Pontes., *et al*. have stated that this variation may be explained by the scant and heterogeneous nature of reported clinical chondrosarcoma series, which usually include different histological tumor variants in the same analysis, and by the advancements in surgery have directly improved survival rates [15].

Conclusion

In the present case, the patient's low socioeconomic status and educational attainment were probably responsible for the neglect of the initial symptoms, which, combined with the unusually aggressive course, allowed this tumor to grow unchecked and become inoperable. This case illustrates the need for health education in low socioeconomic groups. The pursuance of local primary health care groups in spreading awareness and dispelling the myth, fear, and social exclusion is necessary even today and age, particularly among the lower socioeconomic and lesser-educated groups of our population.

Competing Interests

None declared.

Funding

None.

Ethical Approval

Not required.

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