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An Unusual Tumor of Tip of Nose

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Abstract

Introduction: Head and neck sarcomas in adults are rare with an incidence of less than 5%. They usually occur in the skull base, maxilla and larynx. Chondrosarcomas are derived from chondrocytes, embryonal rests, or mesenchymal cells. With only 2 reported cases, chondrosarcomas of the nasal tip is fairly out of the common. We hereby report an uncommon case of grade I chondrosarcoma of alar cartilage of nose.

Case Report: A 72-year-old woman, presented with a history of swelling over nasal tip for 15 years. It gradually increased in size over a period of 6 months. Fine Needle Aspiration Cytology from the lump was suggestive of chondroid syringoma. Capsular dissection of the mass was done along with primary closure of the wound. On histopathological examination it was diagnosed to be a case of grade I chondosarcoma with no tumor deposit in the resection margins.

Discussion: The primary challenge was to differentiate grade I chondrosarcoma from enchondroma. In our case the tumor revealed infiltrative properties and cytological atypia on microscopic examination. Further immunohistochemistry for S100 was positive which confirmed the presence of chondrocytes. Hence, we arrived at a diagnosis of grade I chondrosarcoma. Closest differential diagnosis of grade III chondrosarcoma is chondroblastic osteosarcoma,which shows osteoid formation.

Conclusion: Chondrosarcoma of nasal tip is very rare, and it is uncommon in females. Surgical resection of the tumor is the mainstay for treatment of chondrosarcoma. Adjuvant therapies may be used in high grade chondrosarcomas. Prognosis of grade I chondrosarcomas is fairly good, but as the grade increases prognosis becomes poor.

Keywords: ***Alar cartilage; Chondrosarcoma; S 100; Surgery***

1. Introduction

In adults head and neck sarcomas are rare with an incidence of less than 5% [1-3]. Osteosarcoma, rhabdomyosarcoma are commonly seen in head neck region [2,3]. Males are more commonly the sufferers of this ailment as compared to females. About 70% of cases have been reported in patients who are in their sixties to seventies [4-6]. Chondrosarcoma is a malignant neoplasm which can arise from mesenchymal cells. These mesenchymal cells can undergo differentiation to form cartilage or bone [6]. The sites of predilection for chondrosarcoma in the head and neck region includes skull base, maxilla and larynx [7,8]. Only 0.1% of chondrosarcoma can arise from the nasal tip [6]. Origin from the alar cartilage of nasal tip is even rarer with only two reported cases [9]. Nasal chondrosarcoma usually presents as a swelling which gradually increases in size. Ollier disease and Maffuci syndrome may give rise to this chondrosarcoma. Some bony lesions like chondromyxoid fibroma, Paget disease of bone and etc. may give rise to this condition. History of previous radiation, use of thorium dioxide has been found in some of the cases [6]. Surgical resection with wide local excision is the treatment of choice [6]. Here we are reporting an unusual case of chondrosarcoma of alar cartilage of nose. The mass was present at the tip of nose for a long time without any relevant past history and it was successfully excised.

2. Case Report

A 72-year-old woman came to the emergency department with complaint of a lump hanging from the nasal tip for 15 years (FIG. A). The lump significantly increased in size over a period of 6 months. She also complained of discomfort and dragging sensation over the nasal tip for 6 months. She did not complain of any pain or itching sensation over the lump. She did not complain of any nasal obstruction or nasal bleeding. There was no cervical lymphadenopathy. Medical history revealed that she has diabetes mellitus which was well under control with oral hypoglycemic drugs.

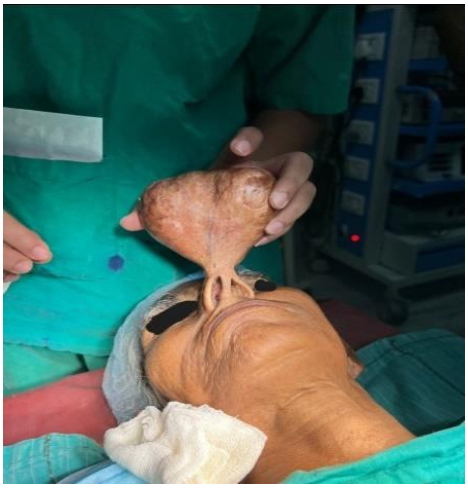

FIG. A

On clinical examination a large bulbous lump was noted over the nasal tip which measured (10×8.5×4) cm (FIG. 1). On palpation the lump was firm in consistency. Skin over the lump was normal, with no redness, no ulceration and no change in temperature. External nares were normal with no obstruction and no deviation of nasal septum.

The fine needle aspiration cytology was suggested to the patient along with other routine investigations like complete blood count, fasting blood sugar, post prandial blood sugar, electrocardiography, and chest x-ray. The complete blood count, fasting blood sugar, post prandial blood sugar, electrocardiography and chest x-ray were within normal limits. Ultrasonography of the mass was done. It showed a 9.7 cm soft tissue mass of heterogenous nature with spotty calcification.

On fine needle aspiration the aspirate was blood mixed particulate. The microscopy of the fine needle aspiration smears showed cellular smears with many epithelial cells which were monomorphic having bland nuclei in a chondromyxoid background (FIG. 4&5). Although mild atypia was appreciated, but there was lack of tumor diathesis in the examined smears. And hence diagnosis of a benign lesion chondroid syringoma was given with an advice of urgent surgical biopsy.

The patient was then referred to the otorhinolaryngology department of our hospital with the above reports. Capsular dissection of the mass with wide local excision was done along with primary closure of the wound. The resected specimen was sent to the department of Pathology for histopathological examination. On gross examination, a skin covered globular tissue piece was received which measured (10×8.5×4) cm (FIG. 2). On cutting open it was whitish homogenous (FIG. 3). Several sections were given from different areas of the tissue piece. Sections were also given from the resection margins. On microscopical examination, the tumor was composed of round to spindle shaped cells alternating with few areas of immature cartilage i.e. biphasic pattern was observed. Infiltrative properties of the tumor were marked on microscopy (FIG. 6-8). The presence of chondrocytes was confirmed by S100 immunostaining (FIG. 9). The diagnosis was consistent with mesenchymal chondrosarcoma (grade I).

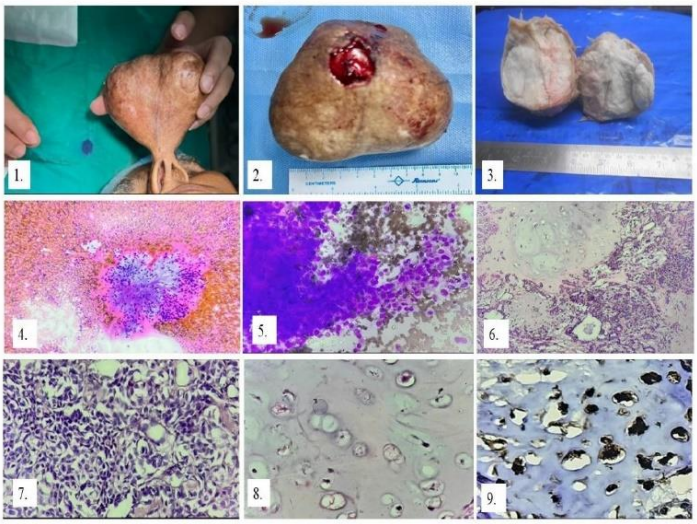

Figure 1: Shows a large bulbous mass arising from nasal tip.
Figure 2: Shows a bulbous mass measuring (10X8.5X4) cm after surgical removal.
Figure 3: Shows cut section of the mass which is whitish homogenous.
Figure 4: Smears showing chondromyxoid background (LG 100X).
Figure 5: Smears showing epithelial cells having mild atypia (LG 400X).
Figure 6: HPE (100X) of mesenchymal chondrosarcoma showing 'biphasic' pattern.
Figure 7: HPE (400X) of mesenchymal chondrosarcoma showing sheets of small round to spindle shaped cells.
Figure 8: HPE (400X) of mesenchymal chondrosarcoma showing cartilage.
Figure 9: HPE (400X) of mesenchymal chondrosarcoma showing chondrocytes positive for S100 immunostain.

3. Discussion

Cartilage or chondroid precursors give rise to chondrosarcomas. Rarely they may arise from tissues having mesenchymal cells but lacking cartilage. Etiology of chondrosarcoma is still a mystery. However, theories stating exposure to iron, aluminum, teflon, asbestos and irradiation as possible causes of chondrosarcoma have been put forward. Secondary chondrosarcomas are seen in about thirty-five per cent of cases. Few examples of diseases which may give rise to chondrosarcomas include Paget's disease, Ollier's disease, Maffucci's syndrome, fibrous dysplasia, solitary, or multiple exostosis [8,10,11]. In our case the patient had no significant past history of having any of these diseases.

Symptoms caused by chondrosarcomas is site dependent [12]. Similarly, skull base chondrosarcomas can produce proptosis, visual loss, vertigo, tinnitus. Sixth cranial nerve palsy is most commonly observed in skull base chondrosarcomas [12,13]. If they are located in the sinonasal tract then they can produce symptoms of mass effects like epistaxis, nasal obstruction [14]. Dysphonia and dyspnea may occur in case of laryngeal involvement [15-16]. In this case the patient had discomfort and dragging sensation as this was hanging from the tip of nose.

Based on microscopical features like nuclear size, atypia, mitotic activity, and degree of cellularity WHO has classified chondrosarcomas are into three grades. Grade I chondrosarcomas have a matrix composed of hyaline cartilage with low cellularity, affecting elderly males in their sixth to seventh decade. Moderately differentiated chondrosarcomas are grade II. High cellularity, mitoses and myxoid matrix are characteristics of a grade III chondrosarcoma [17]. In this case it was a grade I Chondrosarcoma as the cellular atypia was mild with occasional mitotic figures.

Histopathologically, chondrosarcomas can be confused with benign entities like chordoma, chondroma and chondromyxoid fibroma [8,12]. Chondroblastic osteosarcoma is a close differential diagnosis of grade III chondrosarcoma [12]. It is extremely difficult to differentiate low grade chondrosarcoma from enchondroma. Enchondromas usually lack infiltrative properties [12]. Enchondromas usually lack cortical destruction and do not show formation of soft tissue mass in a radiograph [9]. Radiological evaluation of the lesion CT and MRI are crucial for differentiating enchondromas from low grade chondrosarcomas. Chordoma is characterised by presence of "physalliphorous" cells. With the help of Immunohistochemistry D2 - 40. Chordoma will be negative for D2- 40 whereas chondrosarcoma will be positive for D2- 40 [18]. Chondroblastic osteosarcomas show osteoid formation whereas grade III chondrosarcomas usually lack it [19]. In this case the diagnosis was made by Hematoxylin and Eosin stain and confirmed by S-100.

The treatment of Chondrosarcoma is Surgery with wide local excision. Due to its radioresistant nature chondrosarcomas are usually managed by surgical resection. Key to a successful surgery in a case of chondrosarcoma is achieving surgical margins free from tumour deposit [20]. Adjuvant therapies are required in advanced disease and unresectable tumors [10]. The prognosis of chondrosarcoma is usually good with five-year survival rate is 70%- 75% [21]. Rates of metastasis to distant sites is usually low [21]. In our case surgical resection was done with wide local excision. No further treatment was given. Two years have passed. The patient is doing well.

4. Conclusion

Chondosarcoma grade I of the nasal tip is very rare. It is a low-grade malignant tumor. Surgical excision is the treatment of choice. Radiotherapy and Chemotherapy usually are not required. The chance of metastasis is also very less.

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