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"Low-Grade Cutaneous Angiosarcoma of Nose Resection and Reconstruction Using Local Flaps,"

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ABSTRACT
Angiosarcomas are malignant neoplasia's of vessels with rapid growth that develop from endothelial cells. They represent 2% of all sarcomas and only 1–4% are located in the aerodigestive tract. Since 1977, only 16 cases have been reported. We present 72-year-old male patient, with a cutaneous over growth on the nose which was smooth in consistency, purplish discoloration andmeasured about 4*4.2*4.2 cm in size. CT scan showed a nonenhanced tumor on the nose. Biopsy and immunohistochemistry wasperformed which revealed an low-grade angiosarcoma. Surgical excision was performed with immediate reconstruction of the nose and the upper lip with bilateral nasolabial flap and para-median forehead flap. Aerodigestive angiosarcomas have a better prognosis than angiosarcomas of other locations due to better cell differentiation and the presence of early symptoms.

I. BACKGROUND

Angiosarcoma (AS), also known as malignant hemangio-endothelioma, is a rare and aggressive malignant tumor of endothelial cells of vascular or lymphatic origin. These tumors typically have a poor prognosis, and they represent less than 1% of all malignant tumors[1] and between 1% to 2% of soft tissue sarcomas [2-4]. About 50% to 52% of all ASs occur in the head and neck[3,5] and are found predominantly on the scalp and face of elderly men [2,5]. A study showed that ASs represented 11.2% of all sarcomas in the head and neck region [2]. AS is extremely rare in the oral cavity and salivary glands, accounting for 1% of all ASs reported in the literature [1,4]. Although the tumor can occur at any location, the most common sites are soft tissue of skin [6], trunk, and limbs[7]. The tumor type is represented more commonly in male patient specially in the 4th and 5th decade of life as suggested by Fanburg-Smith et al.[8]

II. CASE PRESENTATION

A 72 years old male patient had reported to the surgical oncology department of kiran hospital. With the chief complain of swelling and overgrowth on the nasal tip along with difficulty in breath through the nose since past 2 years. It was a well defined swelling about 4*4*4 cm in size seen on the nasal tip extending to the all both side {bilaterally}and inferiorly extending till the upper lip clinically. The overgrowth is significant thus leading to compression of bilateral nostril with no bleeding or oozing of seen. On palpation the overgrowth was soft, non-erythematous, immobile, non fluctuant. The lymph nodeappeared normal on the clinical examination. All the further investing were carried out for the patient including the lab reports and the blood reports that were normal for the patient and the vitals were noted to be stable. The radiologic examination for the patient included the MRI for the paranasal sinuses the paranasal examination stated that a lesion size 4.5*4.2*4.7 cm in size, ill-defined exophytic fungating heterogenous soft tissue growth involving nose middle third of the upper lip with demarcated invasion in the nasal cartilage the lesion extension shown approximately to the upper alveolus of maxilla the nasal floor and the palate seemed to be clear and CT and CECT for the neck stating and to make the bony correlation of the extension of the tumor indicated that malignant erosive lesion seen affecting anterior part of cartilaginous segment of nasal septum, tip region and right nostril as described. The biopsy for the patient was carried out the and histopathology study concluded as angiosarcoma the further immune-histochemistry study stated **low grade cutaneous angiosarcoma.**

Post clinical examination trail the patient was admitted to the in-patient department of the hospital and along with the subordinate surgeons and the anaesthesiologist the surgical intervention was decided and carried out. The resection was carried out with clear surgical margins in such a way that the nasal tip along with the upper lip, bilateral ala and the nasal septum were resected. The defect reconstruction was done using thea bilateral nasolabial flap and a median forehead flap. During the reconstruction the bilateral nasal flap reconstructed bilateral ala and the nasal tip was formed by the approximation of the forehead flap and lateral the nasolabial flaps. The lip was reconstructed by fusion of the left and right nasolabial flap. Post-surgery the patient was admitted in the ICU for 24 hours and was under continuous surveillance for next 7 days.

III. DISCUSSION

The diagnosis of angiosarcoma is challenging due to the rareness of this tumor, the absence of consistent therapeutic guidelines and its poor prognosis. Therefore, a thorough medical history, a concise physical examination and clinical knowledge are necessary. Cutaneous angiosarcoma is typically a malignancy of elderly males[9], the nose is a rare location and can be easily mistaken for a benign lesion due to its variable presentations[10]. Different clinical pictures have been described in the literature with multiple reports of angiosarcoma mimicking rhinophyma, rosacea, haemangiomas, contact dermatitis and even sebaceous cyst[11]. The lesions are flat or raised nodular, macules and papules of different sizes these feature can lead to a misdiagnosis [10]. Angiosarcomas are known to grow quickly in a centrifugal pattern and subcutaneous extension[12] and this confusion can also delay the diagnosis and treatment, probably explaining in part its poor prognosis. The immunohistochemical markers play a crucial role in the diagnosis of the tumor

Possible immunohistochemical markers for angiosarcoma include ulex europaeus 1 lectin (sensitive marker; used in conjunction with epithelial membrane antigen and cytokeratin to exclude epithelial tumor); factor VIII antigen (highly specific; low detection sensitivity); CD34 cells (highly sensitive; stains dermal dendrocytes, sweat gland basement membrane, and hematopoietic progenitor cells); and CD31 cells (highly sensitive; good specificity).[13] Several differential diagnoses should be considered on pathology.[14] Unlike benign vascular lesions, the well-differentiated angiomatous areas in angiosarcoma display cytologic atypia; multilayering, papillary structures; and irregular anastomosing blood vessels. The optimal treatment of cutaneous angiosarcoma has not been defined.[15] Generally, radical surgery and postoperative radiotherapy are advocated to treat patients with these tumors, surgical excision of the tumor is the best treatment modality and the first line of therapy. There are no strict guidelines concerning the width of surgical margins but clear margins in the anatomopathological examination ensure lower frequency of recurrence. Here in the case it was detected as the low grade angiosarcoma of the nasal soft tissue thus leading to resection of the tumour and followed by immediate reconstruction , Nose reconstruction and upper lip remains challenging as Its goal is both aesthetic and functional. The three-dimensional structure of the nose and lip needs to be reconstructed depending on the location, the depth, size and shape of the nasal defect. The forehead flap remains the gold standard of nasal soft tissue reconstruction providing excellent results[16].Full-thickness reconstruction of the subtotal upper lip defect and the restoration of the function and appearance of the columella has been done using modified bilateral nasolabial flaps [17].

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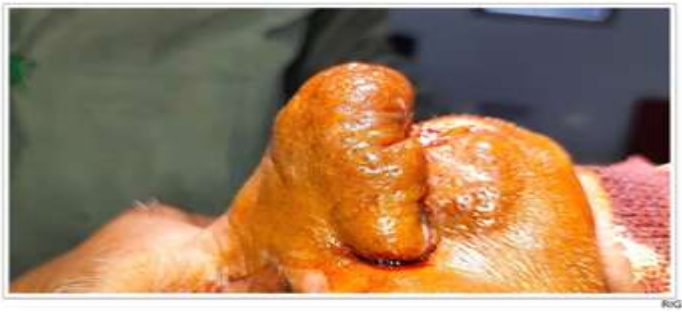
Clinical representation

Clinical pictures

Inferior surface showing the nasal tip and the obstruction



RIGHT SIDE

Incision marking

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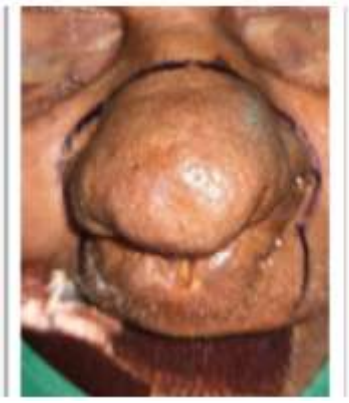
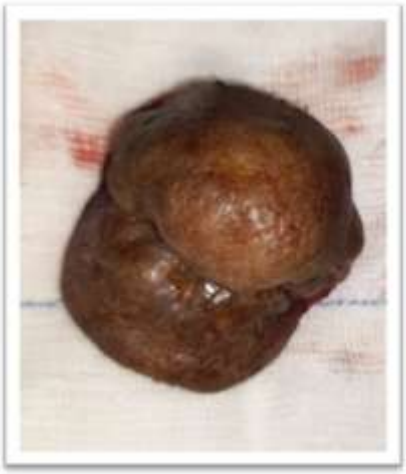
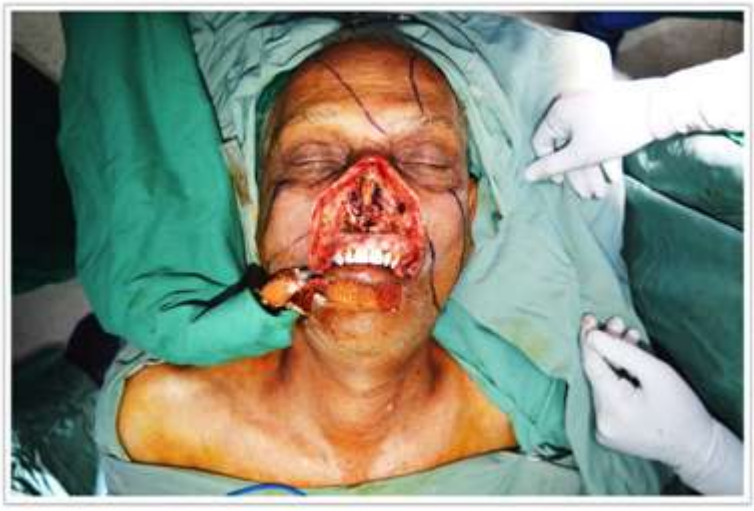


Resected specimen



Bilateral marking for nasolabial flap and paramedian forehead flap



Reconstrution done

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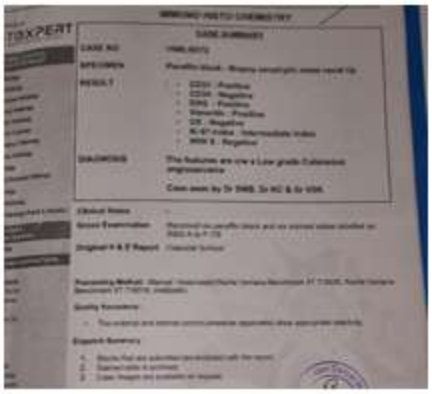
HISTOPATHOLOGY
CASE SUMMARY
CASE NO
SPECIMEN
DIAGNOSIS
Low grade cutaneous Angiosarcoma, nasal tip
ADVICE / COMMENT IHC stains for final diagnosis.
ENCLOSED :- IHC report.of 19 MLJ 8573 confirming a low grade cutaneous angiosarcoma.
Clinical Notes
Gross Examination
Grossed by
Microscopy
Text Performed by


EXPERT
CASE SUMMARY
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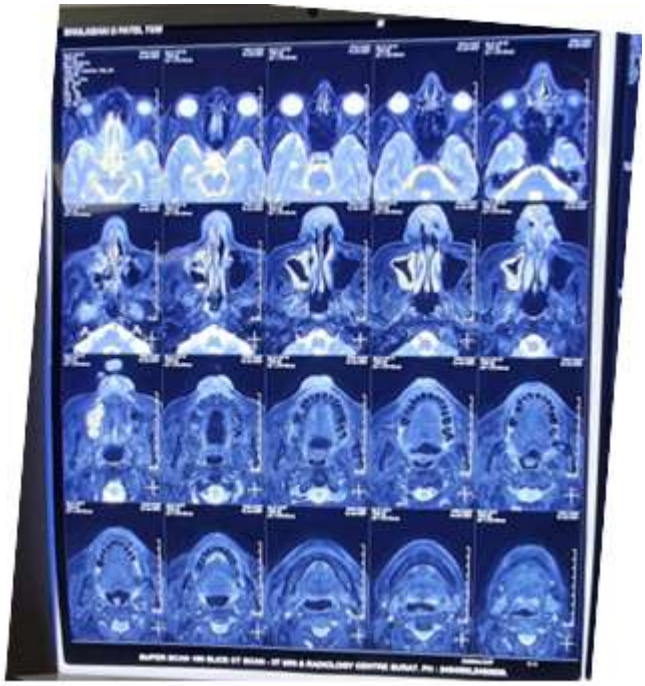
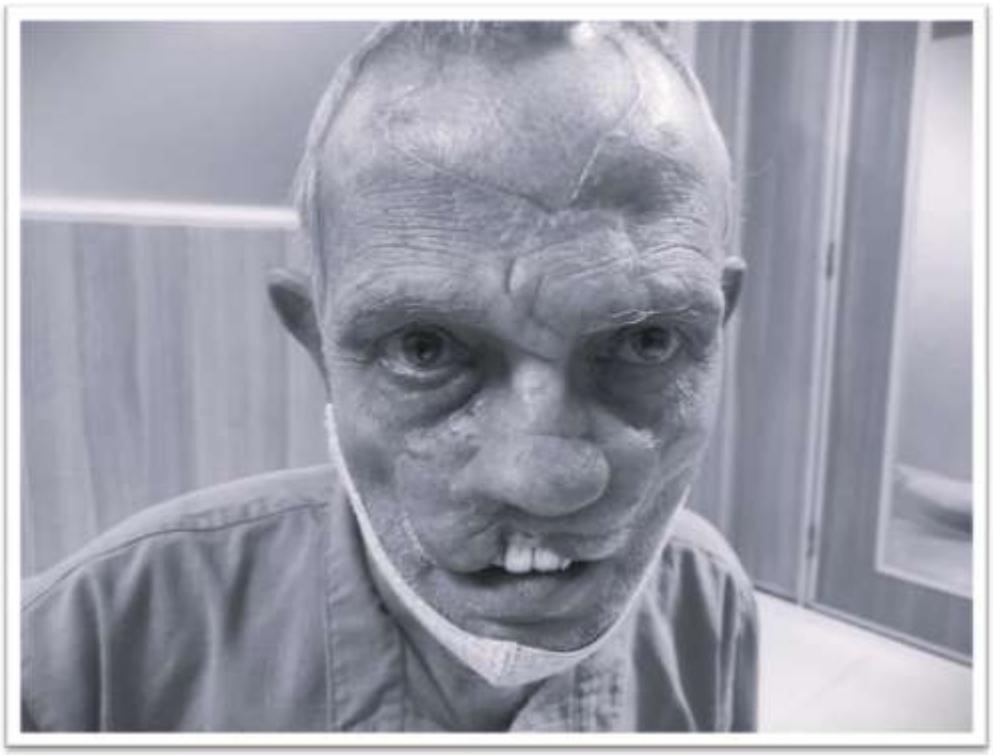
SPECIMEN
RESULT
DIAGNOSIS
Clinical Notes
Gross Examination
Original H & E Report
Immunohistochemistry Summary

Histopathology and histo chemistry reports



radiographic images



post operative image with 6 month follow up