



Giant Fungating Cutaneous Myxoma of the Head and Neck: An Unusual Presentation

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Authors' contributions

This work was carried out in collaboration among all authors. Authors AD, KRI, OWO, TAO, SSM designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors JHS, BRO, AMF, IAA, managed the analyses of the study. Authors MA, OCJ NPA managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Introduction: Virchow first describes Myxomas in 1871; they are benign tumors of primitive indifferent mesenchyme that have preference for the cardiac muscles, genitourinary tract, gastrointestinal tract, liver and spleen. The rarity of the tumor, especially in the head and neck region makes us to report this case.

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Case Report: A 16-year-old male secondary student who presented with a 9-year history of painless progressive submandibular swelling, no extension to the mouth. Examination revealed a huge fungating submandibular mass, 20 cm x 16 cm x10 cm in dimension, mobile, non-tender and firm in consistency. Other findings were unremarkable, and routine hematological and biochemical tests were all within normal limits. He subsequently had excision of the mass (weighs 950 g) under general anesthesia via an elective tracheostomy with split thickness skin grafting of the exposed strap muscles. Histopathological report revealed cutaneous myxoma.

Conclusion: Cutaneous myxoma of the Head and Neck region is presented for its rarity, and although is a histologically benign neoplasm, treatment is extremely challenging with high incidence of recurrence.

Keywords: Cutaneous myxoma; head and neck; benign; submandibular.

1. INTRODUCTION

Myxomas of soft tissue occur in skeletal muscle, subcutaneous tissue, fascial planes, and neurovascular sheaths and tend to arise in the extremities [1,2]. In the head and neck region, the myxomas occur normally in the mandible, maxilla and soft tissues of the face and neck, other sites include the nasal cavity, parotid gland, and oral mucosa [3,4,5]. The tumor was first described by Virchow [6] in 1871, when he described tumors that histologically resembled the mucinous tissue of the umbilical cord. Extramuscular soft tissue myxomas in the cervical region are uncommon and only few cases are described in the English literature [7]. In a study of 49 patients carried out by stout [4], a diagnostic criterion was reported that myxoma is a true mesenchymal neoplasm formed exclusively from undifferentiated cells in a mucoid stroma and does not metastasize. It is not uncommon to have association with Carney complex (CNC) which is an autosomal dominant disorder and characterized by myxomas, spotty pigmentation, endocrine over activity [8] and mutations in the protein kinase AR1 alpha (PRKAR1A) gene [9]. We therefore present an unusual case of a giant fungating cutaneous myxoma of the head and neck.

2. CASE REPORT

A 16 year old male student who presented for the first time to the referral health facility with a 9-year history of painless progressive submandibular swelling, no voice change, difficulty in breathing, dysphagia or extension to the mouth. Examination revealed a huge fungating submandibular mass, 20 cm x 16 cm x10 cm in dimension, mobile, non-tender and firm in consistency. Fig. 1. No palpable lymphadenopathy observed. Other findings were unremarkable, and routine hematological and biochemical tests were all within normal limits.

Surface echocardiogram showed no cardiac lesion. The poor socioeconomic status of the patient with no health insurance coverage limits his access to advanced imaging study like Magnetic resonance imaging of the Neck. Preoperative neck mass biopsy report was in keeping with myxoma. He subsequently had excision of the mass with wide surgical margins under general anesthesia via an elective tracheostomy due to difficult intubation with split thickness skin grafting of the exposed strap muscles. The lesion weighs 950 g and was sent for Histopathological analysis, which shows mesenchymal lesion that is composed of proliferating stellate and globular myxoma cells with abundant eosinophilic cytoplasm, indistinct cell borders, oval nucleus with open chromatin and indistinct nuclei, Abundant mucopolysaccharide (myxoid) ground substance containing chondroitin sulfate and hyaluronic acid. There are delicate vascular network [Fig. 3]. This confirmed cutaneous myxoma. In the postoperative period patient was successfully decanulated and four weeks after excision patient was discharged from hospital. One-year follow up was uneventful.



Fig. 1. Preoperative clinical photograph showing the huge fungating tumor

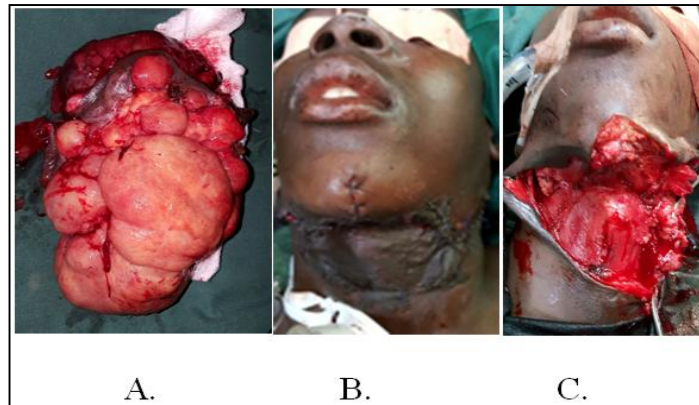


Fig. 2. Intraoperative clinical photographs showing: The excised tumour [A]; Split thickness skin graft [B]; and the wound defect before grafting [C]

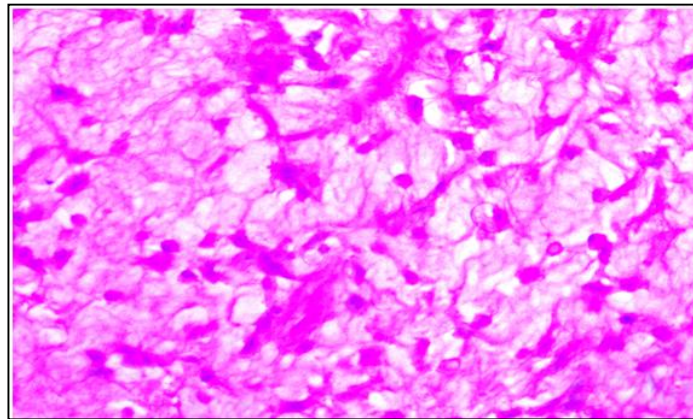


Fig. 3. Photomicrogram of myxoma (H&E, Mag x 100)



Fig. 4. Postoperative clinical photograph showing: [A]. Healed scar at One-year follow up. [B]. Four weeks post excision at discharge from Hospital. And [C.] five days post excision with stoma (white arrow) after decanulation

3. DISCUSSION

The etiology myxoma remains unknown. Documented findings from similar study agree

that myxomas are derived from a primitive embryonal mesenchyme or fibroblasts able to produce mucopolysaccharides in abundance [10]. The most common appearing site of the

tumor is the heart, followed by the thigh and shoulder [11], with infrequent occurrence in the head and neck region [12]. Myxomas commonly present as a slowly growing, painless mass [3] as seen in our patient who had a painless and slowly growing submandibular mass of 9 years duration [Fig. 1]. Myxomas of the head and neck occur with equal frequency in men and women and tend to occur in the third and fourth decades of life, although individuals of all ages can be affected [4,13-14]. In the review of concept of myxoma by Allen [13] in 2000, soft tissue myxomas are classified into intramuscular myxoma, juxta-articular myxoma, superficial angiomyxoma, aggressive angiomyxoma, and Nervous sheath myxoma. The cutaneous myxoma also known as superficial angiomyxoma as seen in our patient appears commonly in the thigh, trunk and head and neck region. Histologically, soft tissue myxoma is composed of spindle or stellate cells and reticulin fibers in abundant mucoid material. The presence of blood vessels and mitosis is infrequent and there is a total absence of mast cells [15,16].

Multiple myxomas of the head and neck that occur in a young patient should alert the clinician to the possibility of a systemic disorder characterized by cardiac and soft tissue myxomas (often of the head and neck), facial and labial skin pigmentation, and endocrine over activity (Cushing's syndrome, acromegaly) [17]. Carney, et al. [18] reported a series of 40 patients with this disorder, which appears to be inherited in an autosomal dominant fashion, reporting 25% of the patients who died of embolization from a cardiac myxoma. The most dreaded presentation of Carney complex is cardiac myxoma that was reported to account for a third of the population. It is pertinent therefore that patients presenting with myxoma symptoms should be screened for cutaneous cardiac lesion [19,20,21]. Our patient had no cardiac lesion on surface echocardiogram.

The treatment of head and neck myxomas is surgical excision with adequate margins to prevent recurrence. Intraoperatively myxoma appears as gelatinous mass of oval or spherical shapes and variable consistency as shown in [Fig. 2A]. Significant ongoing debate has centered on the best mode of surgical therapy, with recommendations range from enucleation or curettage [22] to extensive surgery with wide margins or radiotherapy [23]. Poor definition of tumor margins is a major cause of incomplete resection and subsequent recurrence [5]. Our

patient had resection with wide surgical margins and has had no tumor recurrence a year after surgery [Fig. 3].

4. CONCLUSION

Head and neck myxomas may present with difficulty in achieving wide surgical margins, but to minimize the rate of recurrence, efforts must be made to attain the best clear margins without compromising vital head and neck structures.

CONSENT

As per international standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard, written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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