Angiomyolipoma of Nasal Cavity - A rare clinical entity

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ABSTRACT

Angiomyolipomas (AML) are benign mesenchymal tumour with varying proportion of matured fat cells, thick walled vessels and smooth muscle cells. Nasal angiomyolipomas are exceedingly rare and usually present as small tumour in middle aged and old men.

This is a case of 86 years old male who presented in the ENT OPD with the swelling in the right nasolabial fold for 2 months. He underwent excision of the mass under general anesthesia and the histopathological examination proved to be angiomyolipoma of nose.

KEY WORDS

Angiomyolipoma, Hamartoma, Histopathology

INTRODUCTION

Angiomyolipoma (AML) is a mesenchymal tumor composed of variable proportions of blood vessels, smooth muscle cells, and adipocytes. Angiomyolipoma is generally a benign tumour composed of a mixture of mature fat, smooth muscle and blood vessels in various proportions and 99% of angiomyolipoma is found in kidneys.[1,2] Angiomyolipoma mostly occurs in the kidneys and rarely in the liver, skin gastrointestinal tract, heart, lungs, spinal cord and oropharynx.[3-7] Nasal angiomyolipomas are exceedingly rare and usually present as small tumors in middleaged or old men. They have characteristics common to other mucosal and cutaneous angiomyolipomas, suggesting that the tumors in these locations may be grouped as mucocutaneous angiomyolipoma and are distinguishable from renal tumors. AML was classified in older literature as hamartomatous lesion, a benign tumour comprising cells of origin, but it is also been found to be associated with other diseases such as neurofibromatosis type 1, polycystic kidney disease and Hippel-Lindau syndrome.

CASE REPORT

An eighty six years male presented in ENT OPD with complaint of swelling in right nasolabial fold for last 2 months. Swelling was insidious in onset and gradually progressive associated with nasal obstruction. There was no history of pain, nasal discharge or previous trauma.

Examination of the mass showed a well circumscribed mass in the right ala of nose. It was globular in shape, about 3 X 3 cm in size, non tender, soft in consistency. Mobile and attached to overlying skin. There was associated narrowing of right nasal cavity. CT Scan revealed isodense lesion at right ala of nose, 3 X 2 X 1.5 cm (Fig. 1 and 2).

The mass was excised under general anaesthesia. The surgical specimen was reddish brown in colour, multilobulated and firm. Histopathological examination revealed greyish white tissue altogether measuring 3 X 2 X 1.5 cm. Section showed lobules of mature adipocytes with eccentrically pushed nucleus and abundant cytoplasm. Also seen were numerous thick and thin walled blood

Figure 1. Preoperative photograph.

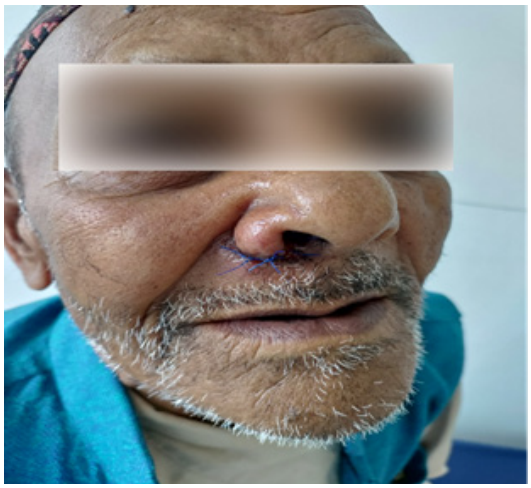

Figure 2. Postoperative photograph

DISCUSSION

Angiomyolipoma is a mesenchymal tumour composed of variable proportions of blood vessels, smooth cells and adipocytes.[8] It was first described by Fitzopahel et al.[9] It belongs to the family of perivascularepitheliod cell tumours which include lymphphangioleiomatosis (LAM), clear cell tumour and simillar tumour that occur in a variety of visceral, cutaneous and soft tissue sites. More than 99% of AML is found in the kidney. Other documented sites are liver, lungs, intestine, oral and nasal cavities and sometimes the skin.

Nasal AML are extremely rare and only few cases have been reported worldwide.[10-12] Cutaneous AML has been reported to be commoner in middle aged male population and rarely in children.[13] Patients with nasal AML are predominantly male and between 28 and 88 years of age. The patient in our case report is of age 86 years old male. The most common location reported is the nasal cavity vestibule. When evaluating a benign mass lesion in the nasal cavity clinically, nasal AML is suggested in the differential diagnosis alongside more common lesions such as nasal polyp, squamous papilloma, juvenile angiofibroma and haematoma. Tumours are reportedly small measuring between 0.8 and 4 cm in contrast to those in the kidney and liver which are larger. The mass in our case was 3 X 2 X 1.5 cm.

vessels, bundle of skeletal muscle fibres, nerve bundles, mild chronic inflammatory cells and areas of haemorrage. Diagnosis of angiomyolipoma was made based on the HPE report (Fig. 3).

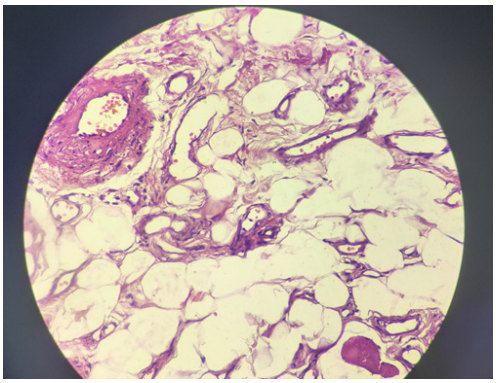

Figure 3. Histopathological image showing adipocytes, muscle fibres, blood vessels.

AML are histologically diagnosed as benign tumours with classical triad of mature thick-walled blood vessel, smooth muscle and mature adipose tissue in different proportion. The histological composition of this tumour comprises vascular, smooth muscles and adipose elements which are intimately admixed qualifying the tumour on inclusion within hamartoma, revealed greyish white tissue altogether measuring 3 X 2 X 1.5 cm. In our case, section showed lobules of mature adipocytes with eccentrically pushed nucleus and abundant cytoplasm. Also seen were numerous thick and thin walled blood vessels, bundle of skeletal muscle fibres, nerve bundles, mild chronic inflammatory cells and areas of haemorrhage.

Hamartoma are benign and self limited lesion and recurrence have not been documented in the nasal AML.[14-16] So, complete surgical excision is the preferred treatment. We also did the complete surgical removal of the nasal mass and the postoperative period was uneventful.

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