Hemangiopericytoma of tongue in a 35 year old male – A rare case report

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Abstract: Hemangiopericytoma is a rare vascular tumour of unknown etiology. It is an uncommon in head & neck. The tongue is the most common site in the oral cavity. The treatment of choice is wide surgical excision. This tumour has a high rate of recurrence and often a malignant transformation.

Keywords: hemangiopericytoma, histopathology, immunohistochemistry.

INTRODUCTION:

Hemangio pericytoma, first described by Stout and Murray in 1942 [1]. It is an uncommon vascular neoplasm characterized by an abnormal proliferation of pericytes (Zimmermann’s pericytes) around thin walled vascular channels and has a predilection for musculoskeletal system [2-4]. This tumor has a highest incidence between third and sixth decade, though it may occur at any age without any predilection for sex. This tumor may behave benignly, being well defined, slow growing, painless and firm, and having a normal overlying mucosa [2, 3, 6]. We are presenting a rare case of a 35 year old male diagnosed as Hemangiopericytoma of tongue on histopathology after planned surgical excision of the tumour.

CASE REPORT:

A 35 year old male came to the outpatient department of the ENT at Rohilkhand Medical College and Hospital, Bareilly (UP), complaining of a slow growing mass on his tongue since last six months. On examination a well-defined pedunculated firm mass of about 3*3 cm size was seen on the tip of the tongue over right side (Fig.1A). As told by the patient, there was no history of trauma and the mass was painless. The overlying mucosa was normal. There were no palpable cervical lymph nodes. The differential diagnosis included a fibroma and granular cell myoblastoma. The lesion was completely excised taking 5 mm margins under general anaesthesia (Fig.1B) and tissue was evaluated histopathologically. The gross features of the tissue were a round, firm, purplish brown mass, and 3.5cm in diameter. Microscopically tissue comprised of both hypocellular areas alternating with hypercellular areas & thin walled branching vessels with gaping sinusoidal spaces (staghorn configuration). Tumour cells are spindled to round with small amount of eosinophilic cytoplasm, bland vesicular nuclei without atypia (Fig.2). Immunohistochemistry showed a positive reaction towards the following markers: bCL2, CD34, and negative towards desmin and CK (Fig.3), yielding the diagnosis of hemangio pericytoma. The margins were negative, therefore no adjuvant treatment given. The patient was followed for 12 months, and no recurrence was seen.

Fig.1-A: showing mass over right side of the tip of the tongue. B: postop after 12 months
DISCUSSION:

The Hemangiopericytoma is uncommon in the head and neck and it represents about 1% of all the vascular tumors [6] and it usually affects adults [4]. Stout & Murray (1942) described 691 cases of vascular tumors, and only nine of them were hemangio pericytomas [1]. Since then, there are approximately 300 cases of hemangio pericytomas described, especially on the trunk and lower limbs [6]. Only 15% to 30% of these tumors are found in the head and neck [7]. At this location, it affects mainly the soft tissue surrounding the oral cavity, sinus tract and meninges and, more rarely, the orbit, parotid gland, skull base and temporal bone [6]. Image studies, such as radiographies, CT scans and angiography are not specific. MRI reveals a solid mass with isodense contrast in T1 [6]. The literature reveals that the tongue is the most common site for the intra oral hemangio pericytoma. The treatment of choice is complete surgical resection of the lesion. Adjuvant radiotherapy and chemotherapy may be indicated in cases in which there is only a partial resection [6]. Though an innocent looking tumor, it has a high recurrence rate. Stout found it to be 28% [5] and Backwinkel 52.2% [2]. Malignant change is also quoted in the literature of 9 to 60% [3, 5, 8-10] but rare in patients treated with complete surgical excision. Most of the patients who had metastases or recurrences were diagnosed after over 40 months of follow up; suggesting a long standing postoperative follow-up for all the patients [7].

REFERENCES: