
CASE REPORT**Glomangioma on Right Forearm - A Rare Case Report***Santosh L. Munde¹, Hemlata T. Kamra^{1*}, Kulwant Singh¹, Swaran Kaur¹, Parveen Kundu¹**¹Department of Pathology, BPS Govt. Medical College, Khanpur Kalan, Sonapat-131001
(Haryana) India***Abstract:**

Glomangioma is very rare lesion in extradigital location. They are usually associated with multiple and familial lesion. Glomangioma as with glomus tumour also presents with hypersensitivity to cold, severe pain and pinpoint tenderness to blunt palpation. Glomus tumours have not been observed in individuals who are less than one year of age (glomus bodies form after that age) or in elderly persons (glomus bodies undergo atrophy and degeneration in elderly persons). Diagnosis is confirmed by histopathological diagnosis but should be differentiated from glomangiosarcoma which shows nuclear atypia and mitotic activity. We here present a case of glomangioma in a 28 years male on right forearm.

Keywords: Glomangioma, Glomus Bodies

Introduction:

Glomus tumours are considered hemangiomas or benign localized tumours of the skin accounting for 1% to 2% of all soft tissue tumours. They originate from the neuromyoarterial plexus: modified smooth muscle cells of the glomus body. Glomus tumours have two forms with the more common solitary variant accounting for 90% of cases and a rarer multiple variant, termed glomangioma, accounting for 10% of cases [1]. The term glomangioma was coined by Bailey in

1935[2]. Glomangiomas are predominant in males, while females (in 70% of cases) are found to have solitary glomus tumours more frequently [3]. Glomangiomas have a predilection for the upper extremities and occasionally are found on the lower extremities, head, and back. They may be slightly larger and less well circumscribed than solitary glomus tumours. We here present a case of glomangioma in a 28 year old male on right forearm.

Case Report:

A 28-years-old male came to orthopaedics OPD with a painful lesion over the right forearm of six months duration. The lesion was single small round reddish blue papule which gradually increased to the present size of 0.5 cm. There was no history of trauma preceding the swelling. The papule was firm in consistency and tender. Excision biopsy revealed a tumour with plenty of vascular channels and a large thrombus (Fig.1). The tumour consisted of sheets of uniform cells with pale cytoplasm; round to ovoid centrally placed punched out nuclei and well defined cell margins (Fig. 2 and 3). The numerous dilated thin walled vascular spaces were surrounded by layers of tumour cells consistent with the diagnosis of glomangioma.

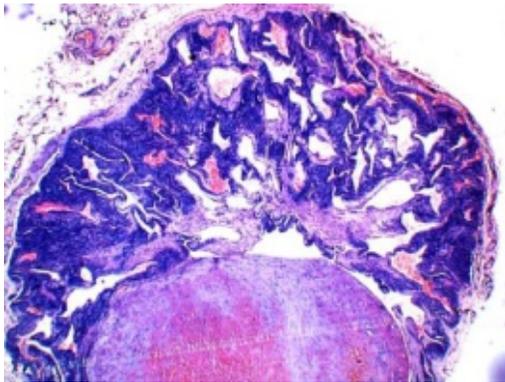


Fig.1: (H&E Stain, 4x) shows Vascular Channels Surrounded by Glomus Cells along with Thrombus

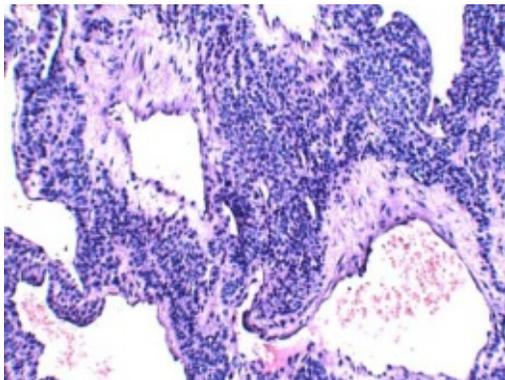


Fig. 2: (H&E stain, 10x) shows Glomus Cells Surrounding Vascular Channels

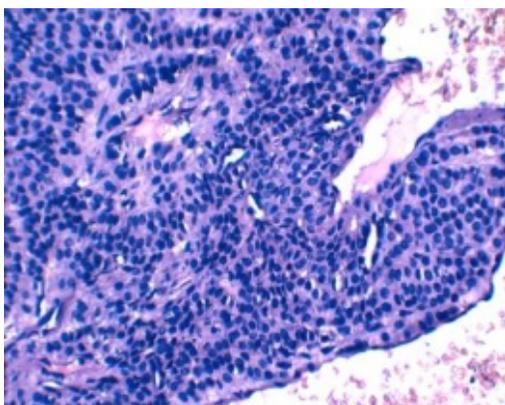


Fig. 3: (H&E stain, 40x) shows Glomus Cells Surrounding Vascular Channels

Discussion:

Typical glomus tumours are subcategorized as "solid glomus tumour", "glomangioma", and "glomangiomyoma" depending on the relative prominence of glomus cells, vascular structures and smooth muscle. In 1924, Masson [4] described the neuromyoarterial glomus, which he later renamed the neurovascular glomus, and its tumours but the term glomangioma was coined by Bailey [2] Glomangiomas are the most common type of glomus tumour in patients with multiple or familial lesions. Most common site of occurrence is in the hands, especially in the distal phalanx. The unusual locations have also been reported such as in the ankle, foot, knee, thigh, hip, stomach, tongue, lung, sella, trachea, and vagina [5]. Extra digital localization of glomus tumour is also rare. Histologically, glomangiomas contain clusters of dilated vascular channels lined by a thin layer of endothelial cells in the dermis or subcutaneous fat. Dilated vascular channels surrounded by glomus cells characterize glomangioma. Immunohistochemically, glomus tumours of all types typically express smooth muscle actin and have abundant pericellular type IV collagen production. H-caldesmon is also positive. Other markers, including desmin, CD34, cytokeratin and S100 protein are usually negative. Glomus cells are small, uniform, rounded cells with a centrally placed, round nucleus and amphophilic to lightly eosinophilic cytoplasm. Each cell is surrounded by basal lamina, seen best on PAS or toluidine blue stains. Occasionally cases show oncocytic or epithelioid change.

Genetic studies have shown that they arise secondary to truncating mutations of the glomulin gene located at 1p21-22. Although glomulin is a normal component of vascular smooth muscle during embryogenesis, it is not yet understood how its functional absence relates to the development of these malformations. Based on analysis of several families with predominantly inherited lesions, four principal germline mutations have been identified. Glomangiomas have been termed malformations. Random post-zygotic mutations may explain both the variation in the number and distribution of lesions in familial cases and the occurrence of non familial cases [6].

On ultrasound the lesion presents as nonspecific solid hypoechoic mass. The high-velocity flow in intratumoural shunt vessels causes this lesion to be hypervascular at color Doppler which is a specific finding for subungual hemangioma. MRI features include intermediate or low signal intensity on T1-weighted images, marked hyperintensity on T2-weighted images, and strong enhancement after the injection of gadolinium-based contrast material. Although MRI is reported to be definitive in subungual glomangioma, it is not specific for extradigital locations [7]. Glomangioma is to be differentiated from the painful tumours of the skin such as eccrine spiradenoma, where two populations of cells and focal ductal differentiation are seen. It should also be distinguished from Blue Rubber Bleb Nevus Syndrome (BRBNS), which is associated with venous malformations on both the skin and

gastrointestinal tract. Cutaneous lesions of BRBNS are blue, soft, and nipplelike, easily compressing and refilling slowly [8]. On the other hand, glomangiomas are noted for a distinct raised, often hyperkeratotic, cobblestonelike appearance and could not be completely emptied by compression [9].

Glomangiomas generally do not extend into deep structures. Histopathologically the differential diagnosis is glomangiosarcoma which can be ruled out on the basis of hyperchromasia, nuclear atypia and mitotic activity.

The treatment of choice for isolated glomangioma is surgical excision. Sclerotherapy with sodium tetradecyl sulphate, polidocinol and hypertonic saline has been reported to be effective in patients with multiple glomangioma located on the extremities. However, sclerotherapy was found to be ineffective in a series of seven patients with large glomangioma [10]. Ablative therapy with argon and carbon dioxide laser is of potential benefit for small superficial lesions.

In this case the lesion is present in an unusual site and could not be diagnosed preoperatively because of unusual location. The patient had presented only with complain of severe pain and he had no other lesion on the body. Therefore lesion was excised. Thus glomangioma should also be considered in differential diagnosis with leiomyoma, eccrine spiradenoma, neuroma, dermatofibroma, angioliipoma and neurilemmoma. Most important is blue red colour of lesion along with pinpoint tenderness to blunt palpation which can help in clinical diagnosis.

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