

Case Report on Glomus Tumor at Extra-Digital Site: A Rare Occurrence

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ABSTRACT

Glomus tumor, also known as Glomangioma, originates in the neuromyoarterial glomus, a normal arteriovenous shunt abundantly supplied with nerve fibres and fulfilling a temperature-regulating function. The classic location of the glomus tumor is the subungual region, but it can occur elsewhere in the skin, soft tissues (particularly in the flexor surface of the arms and about the knee), nerves, stomach, nasal cavity, and trachea. It has also been reported in the sacrococcygeal region, arising from the coccygeal body (glomus coccygeum) and associated with coccydynia, but there is some question as to whether this is a true neoplasm or simply a normal structure.

KEYWORDS: Arteriovenous Shunt, Coccydynia, Glomangioma, Glomus Tumor, Neuromyoarterial Glomus, Subungual

INTRODUCTION

Glomus tumor is a neoplasm which arises from modified smooth muscle cells of the normal glomus body. The glomus body is a specialized form of arteriovenous anastomosis which is involved in temperature regulation.

CASE REPORT

A 45 year-old female patient presented to surgery outpatient department with complaints of a painful mass for 20 years on the external surface of right forearm.

Examination

A small purple coloured nodule of size 1×1 cms was noted on the external surface of right forearm which was very tender on palpation.

Surgery

The painful nodule was excised completely under short general anaesthesia. The tissue was sent for histopathological examination.

Pathology

Grossly the specimen was slightly purple in colour of size 0.5×0.5 cms.

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Histopathological findings revealed the lesions to be Glomus tumor consisting of blood vessels lined by normal endothelial cells and surrounded by a solid proliferation of round or cuboidal 'epithelioid' cells with perfectly round nuclei and acidophilic cytoplasm (Figure No.1,2)

DISCUSSION

The glomus tumor usually presents as a purple nodule, often in a subungual location. These lesions are characteristically painful, a sensation that either arises spontaneously or is elicited by pressure. A variant of this tumor, glomangioma, is often solitary but may be multiple and can be inherited as an autosomal dominant trait. These lesions occur on the trunk or extremities and are most often painless.

The glomus tumor is well circumscribed or encapsulated and is composed of rather solid aggregates of glomus cells surrounding relatively small vessels, whereas glomangiomas are poorly circumscribed and feature prominent vessels surrounded by only a few layers of glomus cells.¹ Immunohistochemistry shows that glomus cells are smooth muscle actin and muscle-specific actin positive and are negative for desmin and markers of endothelial cells. Multiple nerve fibers can be identified in solitary glomus tumors. Most glomus tumors are benign, but atypical examples exist. The study of atypical glomus tumors by Folpe et al. resulted in a classification scheme that categorizes these lesions as follows: malignant glomus tumor, symplastic glomus tumor, glomus tumor of uncertain malignant potential, and glomangiomas.² Malignant tumors were considered those with deep location and size greater than 2 cm, or atypical mitotic figures, or moderate to high nuclear grade and five or more mitotic figures per 50 high-power fields. Symplastic glomus tumors featured high

nuclear grade but no other malignant features. Metastasis occurred only in the group of tumors fulfilling criteria for malignancy.

Various microscopic types of glomus tumor have been recognized: Three important types are solid, angiomatous, and myxoid variety of glomus tumor. The solid type has to be differentiated from sweat gland tumor, melanocytic nevus, or metastatic carcinoma especially when tumour cells are epithelioid type. The angiomatous type can have a diffuse quality, in the sense of looking like an angiomatosis with an increased number of glomus cells in the vessel wall. An oncocytic variant of glomus tumor has also been described, in which the cytoplasm of the glomus cells is packed with mitochondria.

Although Glomus tumors are commonly situated at subungual locations but they are rarely also present at extra-digital sites also, so Glomus tumor must be considered as a differential diagnosis in patients presenting with purple coloured, small, compressible, soft, extremely painful nodule at sites other than subungual region of digits.⁴

When glomangioma exhibits prominent hemangiopericytoma-like vascular features, they are referred to as glomangiopericytoma. There is variation in amount of stromal tissue in glomus tumors & in certain cases there is prominent fibrosis with myxoid changes. Glomus tumors are painful lesions due to entrapment of myelinated nerve fibres. Majority of glomus tumors are benign and cured by excision. Recurrence after local excision is seen in 10% of cases.⁵

Malignant glomus tumors are rare. They are characterized by tumor size of more than 2cms with atypical mitotic figures and marked nuclear atypia.⁶

IMMUNOHISTOCHEMISTRY OF GLOMUS TUMORS

The tumor cells reveal immunopositivity for vimentin, smooth muscle actin, caldesmon and Calponin.

DIFFERENTIAL DIAGNOSIS OF GLOMUS TUMORS

- Eccrine spiradenoma
- Hemangiopericytoma
- Nodular hidradenoma

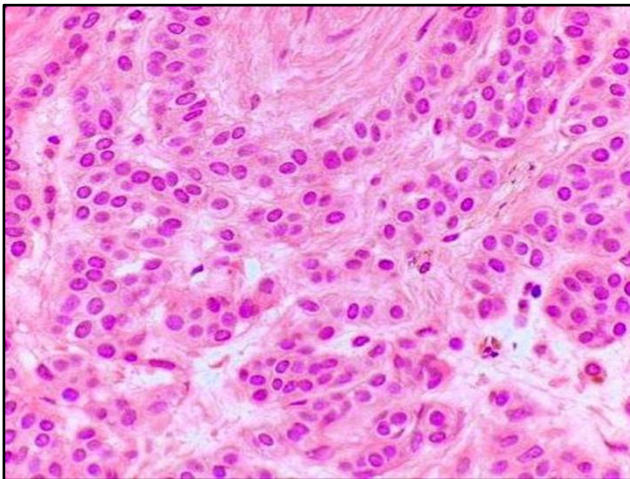


Figure No.1: Showing round to oval cells of uniform size and shape

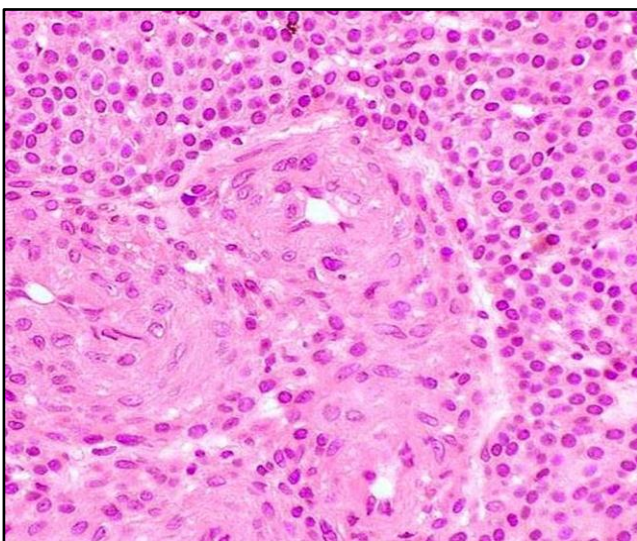


Figure No.2: Higher magnification, showing round cells in clusters

CONCLUSION

Glomus tumours are commonly located in the subungual region of finger. Other sites include palm, wrist, forearm, foot and tip of the spine. It's a Solitary painful lesion composed of glomus cells which are round, regular shaped with a sharply punched out rounded nucleus.

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