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Authors

Rao, Raghavendra
Tejasvi, Bobbili Reddy
Pai, Kanthilatha

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Photo Vignette

Purplish tender nodule on the arm

Raghavendra Rao, Bobbili Reddy Tejasvi, Kanthilatha Pai

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Kasturba Medical College, Manipal University, India

Correspondence:

Raghavendra Rao
Department of Dermatology
Kasturba Medical College, Manipal University, India
jennyrao1@yahoo.com

Abstract

We present a patient with a painful glomus tumor of the right upper arm.

Case synopsis

A 74-year-old man presented with a painful nodule over the left arm of 30 years duration. Pain was intermittent initially, burning in nature, and aggravated by friction with clothes. He reported worsening of pain over the past two years. Examination revealed a purplish, tender nodule measuring 1.0 x 0.5cm over the posterior aspect of left upper arm (Figure1). His baseline hematological and biochemical parameters were normal. Ultrasound examination revealed a well-defined hypo-echoic mass showing significant internal vascularity, predominantly in the subcutaneous plane, with no infiltration into underlying triceps muscle.

An excisional biopsy was done and the entire nodule was sent for histopathological examination. Histopathology revealed a well-circumscribed tumor in the dermis composed of round cells interspersed with numerous blood vessels (Figures 2 & 3)



Figure 1. Purplish nodule on the left arm

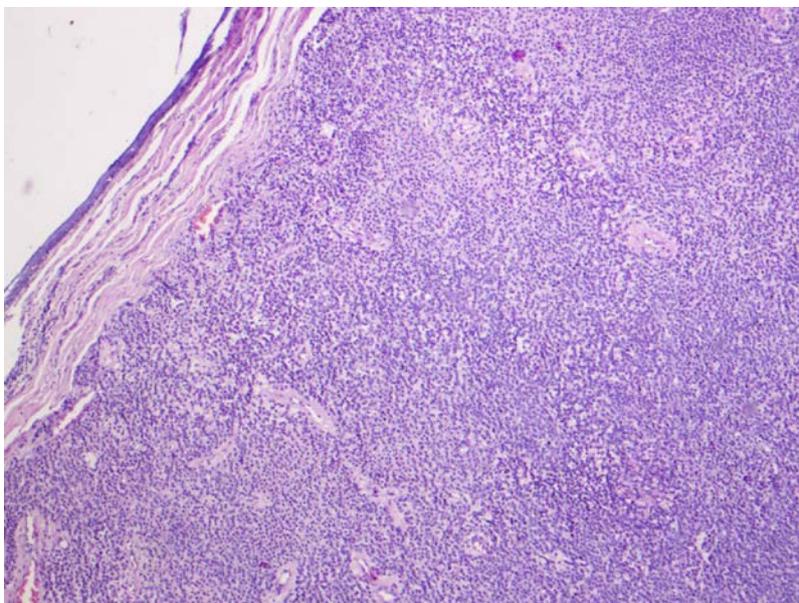


Figure 2. A well-circumscribed tumor in dermis composed of uniform round cells (Haemotoxylin and eosin, X50)

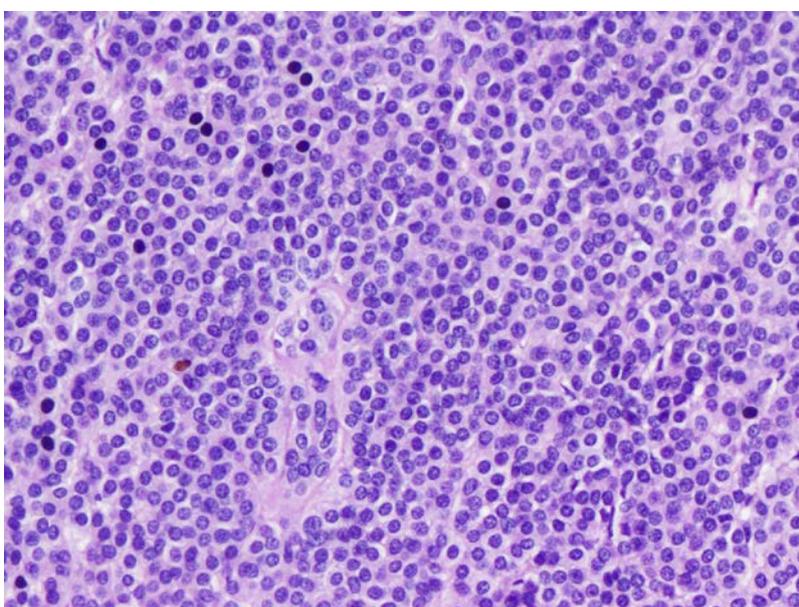


Figure 3. Sheets of monotonous tumor cells having round regular nuclei with finely dispersed chromatin interspersed with numerous blood vessels (Haemotoxylin and eosin, X400)

Discussion

Glomus tumour (GT) is a benign mesenchymal neoplasm arising from the glomus bodies. A glomus body is a specialized form of arteriovenous anastomosis, which plays a key role in thermoregulation. GT accounts for less than 2% of soft tissue tumors; it usually occurs in areas rich in glomus bodies such as the subungual regions of digits or the deep dermis of the palm, wrist, forearm, and foot [1].

GT are categorized into two forms, solitary and multiple. Solitary GTs account for 90 percent of all GTs and are seen more commonly in adult women [2]. The tumors present as deep blue or purple colored papulo-nodules in the hand, rarely exceeding 1.0 cm in its greatest dimension. GT is characterized by a classic triad of pain, pinpoint tenderness with blunt palpation, and hypersensitivity to cold. Concurrence of these symptoms has been shown to enable a clinical diagnosis in more than 90% of cases [3]. On the other hand, multiple GTs are hereditary; they are seen more often in young people and are usually painless, although painful and painless lesions may coexist in the same patient. They are seen more frequent on upper limbs and have an equal sex distribution. Clinical diagnosis of GT remains a challenge for both solitary and multiple lesions; the mean duration of symptoms prior to correct diagnosis is reported to be between 7 to 11 years [3].

Extradigital GT accounts for less than 30% of all GTs and are more difficult to diagnose than digital GTs, resulting in delayed diagnosis or misdiagnosis [4]. These tumors are more common in older males. The incidence of pain and cold intolerance are significantly lower in patients with extradigital tumors. Lee et al have found that extradigital GTs are most frequently seen on the upper arm and forearm [4].

GTs are typically composed of three components: glomus cells, blood vessels, and smooth muscle cells. The glomus cells appear as round to cuboidal cells with round/oval punched-out nuclei and slightly eosinophilic cytoplasm. Based on the relative dominance of cell type, GTs are subcategorized as solid glomus tumor (with poor vasculature and scant smooth muscle component), glomangioma (with prominent vascular component), or glomangiomyoma (with prominent vascular and smooth muscle components). Solid glomus tumor is the most common variant (75%) followed by glomangioma (20%) and glomangiomyoma (5%) [1]. The glomangioma subtype appears to be more frequently encountered at extradigital sites [4]. Recently, MRI has emerged as a sensitive imaging modality that can be very useful in the evaluation of GT, especially when they are small and non-pigmented [5].

Malignant transformation of GTs (i.e., glomangiosarcomas) is exceedingly rare and accounts for less than 1% of all GT cases [6]. Cutaneous glomangiosarcomas may originate from the preexisting GT or may arise de novo; rarely it may represent cutaneous metastases from an internal glomangiosarcoma. Criteria have been suggested for defining malignancy in glomus tumors and estimating the risk of recurrence and metastasis. These include: deep location, size > 20 mm, atypical mitotic figures, the combination of moderate to high nuclear grade, and mitotic activity > 5 mitoses per 50HPF. A much more favorable prognosis has also been noted for internal malignant glomus tumors compared with those found at deep peripheral sites [7].

Complete excision is thought to be curative for solitary lesions, resulting in resolution of all symptoms. Recurrences following excision of solitary tumors have been reported in 12% to 33% of cases. Alternatively, hypertonic saline injection, flash lamp tunable dye laser, and sclerotherapy have also been advocated [3].

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