

Juxta Axillary Granular Cell Tumor

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Abstract: Granular cell tumors or Abrikossoff tumors are very rare, benign and unique tumors in the vast majority of cases, described for the first time in 1926, and can mimicking carcinoma clinically and radiologically. The common sites are the oral cavity, then the subcutaneous tissues of the head and neck and the breasts. The diagnosis remains histopathologic. Its local aggressiveness and its potential for recurrence are related to the quality of the first surgical excision. The evolution of the granular cell tumor is often favorable. We report a new observation of juxta axillary granular cell tumor. Our goal is to discuss the diagnosis and therapeutic features of this rare tumor.

Keywords: granular cell tumor, immunohistochemistry, histopathology, surgical resection.

INTRODUCTION

Granular cell tumor (GCT) is a rare lesion, benign and unique in majority of cases. It's a bewildering lesion for clinicians and radiologists and pathologists owing to unusual presentation simulating a breast carcinoma. Prognosis is good following complete surgical excision. We report here a new case of juxta-axillary granular cell tumor. We also go to discuss the diagnosis particularities and managing methods of this exceptional lesion.

CASE PRESENTATION

A 45 year old premenopausal woman, with no medical history consults in our unit for a painless mass in the axillary extension discovered by self-palpation which gradually increasing in size. There was no staining skin or increasing in size during menstruations.

Physical examination revealed a right rounded juxta axillary lesion measuring 3cmx 2cm, it was mobile and well limited, and no adherent to superficial or deep layers. Furthermore, there were no inflammatory signs and no lymphadenopathy palpable in the right axilla .the left breast and axilla were both normal with no diseases. Tumor classification was T2 N0 Mx.

Mammography showed hydric aspect of the opacity in the right axilla. (Figure1). Ultrasonography revealed a solid juxta axillary nodule with irregular

border, hypo echoic with echoic halo, with a perpendicular maximum diameter of 26 mm and acoustic shadow (Figure 2).

A biopsy was performed followed by histologic examination showed a proliferation of big size cells well-arranged, carrying an eosinophilic abundant granular cytoplasm. The nuclei were regular and rounded without atypia or mitotic figures. The stroma was fibrous and inflammatory with no suspect lesion or necrosis (Figure3). Immunohistochemically staining showed an immunoreaction to the S-100 protein and none to cytokeratin AE1/AE3, the cells also expressed the CD68 (Figure4).In front of this typical aspect the diagnosis of granular cell tumor was made. A wide local excision was performed and the pathology report confirms the diagnosis. The margins were negatives. Two years after surgery the patient is alive and well with no recurrence during the follow up.

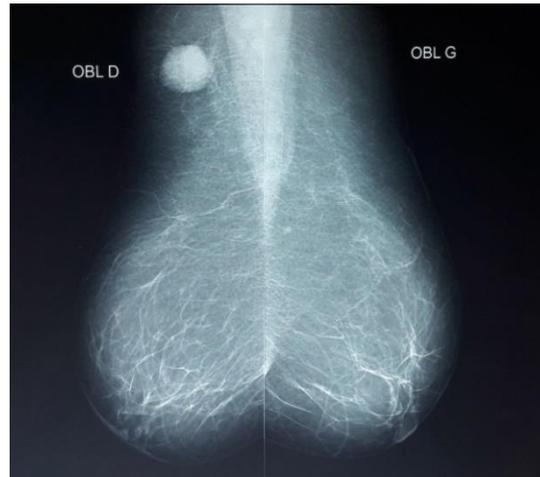


Fig-1: Mammography shows opacity in the right axilla with hydric tonality

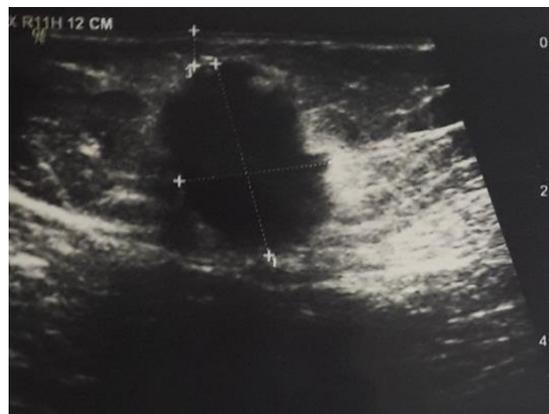


Fig-2: ultrasound view showing right juxta axillary lesion BIRADS 4: the nodule is hypo echoic nonhomogeneous and irregular measuring 26x21 mm

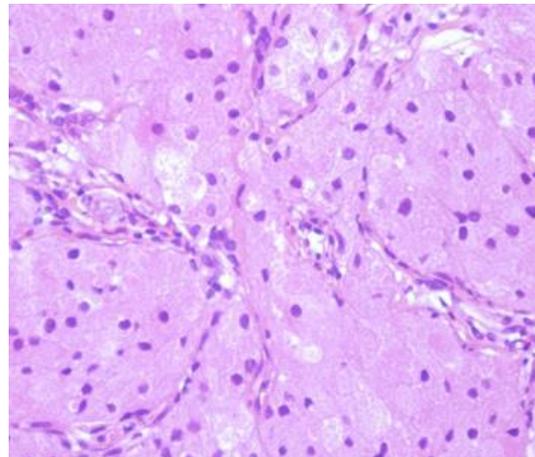


Fig-3: Section of sheets of large polygonal cells with abundant granular eosinophilic cytoplasm

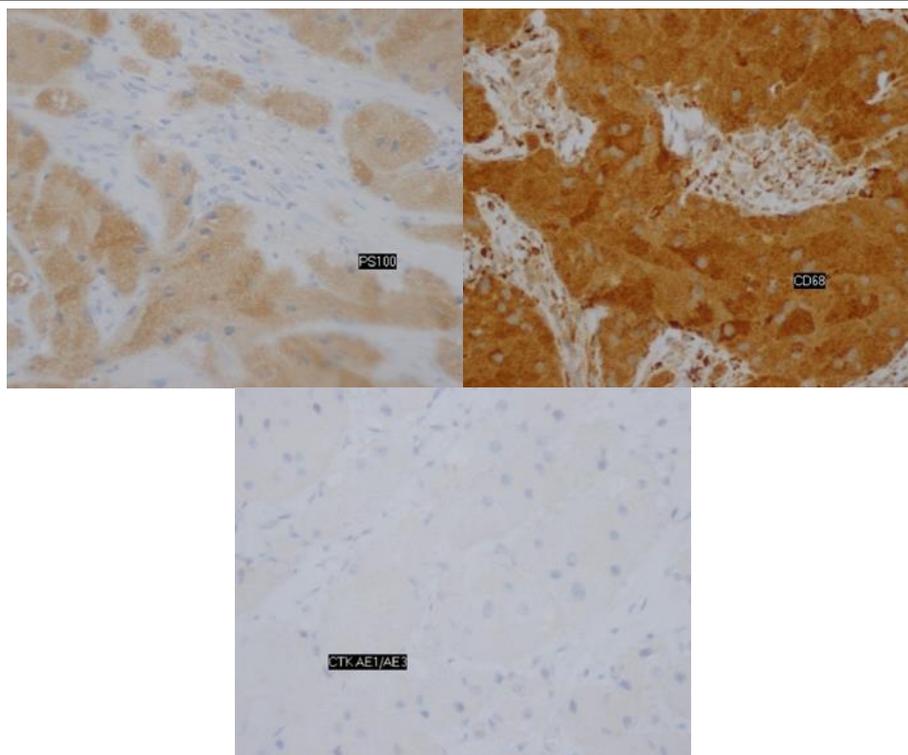


Fig-4: a- PAS-100 staining shows positive activity on the granular cells

b- Positive immunoreaction to CD-68.

c- Negative immunoreaction to cytokeratin AE1/AE3



Fig-5: Macroscopic view of granular tumor measuring 27x25mm

DISCUSSION

Granular cell tumor (GCT) is habitually benign and rare [1], the first description in breast was made by Abrikossov in 1926 by the term of granulo-cellular myoblastoma. In the first time, it was considered as a myogenic lesion of female breast, recent studies opt for schwann cells origin confirmed by immunohistochemistry [2]. Granular cell tumor is omnipresent lesion, affecting essentially soft tissues mainly oral cavity. Other localizations have been documented in digestive tract, bronchial tree, parotid and thyroid glands, kidney and orbit [3].

Clinically, GCT is presented as a painless mass, sharply circumscribed and mobile. Furthermore some lesions are hard in consistency, irregular and

adherent to deep layers, simulating easily a carcinoma. Multifocal presence is reported in 5, 4% to 17, 6% of cases [4, 5]. In our observation, the tumor was unifocal grossly round, mobile, well limited, with a maximum diameter of 3 cm.

Radiological diagnosis of GCT seems to be very difficult owing to similarity with carcinoma and misleading benign aspect without specific signs. Typical aspect in mammography shows a stellar mass with central density and acoustic shadow [6, 8]. This description was found in our case.

Final diagnosis of GCT is made with histopathological examination; we can proceed to micro biopsy or percutaneous biopsy ultrasound guided

allowing good sampling before tumor resection and radical treatment can be avoided [7]. In our case, histologic examination of percutaneous biopsy confirm the diagnosis before total resection of tumor.

Macroscopic aspect of tumor is generally hard lesion in consistency, homogenous, sharply circumscribed, and greyish-white or yellowish, measuring less than 3 cm, but a size of 6 cm was reported [9]. In our patient, the tumor was hard, homogeneous, well limited, yellowish, and measuring 27 mm in the major axis.

The microscopic inspection showed nets of polygonal cells with eosinophilic, abundant granular cytoplasm, the nuclei is small, round and centralized with no atypia. The whole is tight in a fibrous stroma mixed to lymphocytes and plasma cells [10] in this study, the typical morphological aspect was sufficient to make the diagnosis of GCT.

Immunohistochemically study of cells is positive for S-100 protein in 100% of cases and for neuron-specific enolase (NSE) in 90%. It is negative for muscle markers (actin, desmin), and estrogen receptors, and moreover for cytokeratin and the epithelial membrane antigen [11, 12]. Furthermore, the cells shows a positive reaction for CD-68, carcino-embryonic antigen and vimentin in some cases reported in literature. In our case, the cells expressed the PS-100 and CD-68 and the reaction was negative against the cytokeratin AE1/AE3.

Treatment of GCT is wide resection; the final diagnosis is made by correct histologic examination of specimen who must seeks the limits of excision and malignancy criteria [13]. There are malignant forms in 1 to 3% of cases, suspected clinically by the size more than 4 cm, and the presence of necrosis and quick clinical and radiological modifications. Therefore, histology highlights many nuclear atypia, necrosis, and lot of mitoses with increased mitotic index ki67.

Surgical excision is standard treatment of GCT, and adequate margins are necessary to exclude any malignant sign [14]. In our case, the margins were negative and the patient has a complete resection.

The prognosis of this benign tumor is excellent following surgical excision with low recurrence. Close clinical and radiological monitoring is required for any resected GCT [15]. Our patient remained free of disease during a 2 years follow-up period.

CONCLUSION

Granular cell tumor is rare entity; it's difficult to identify owing to clinical and radiologic similarity with carcinoma. Histopathology confirms the diagnosis, and wide excision is the standard treatment. Prognosis

is good, however a close clinical and radiological monitoring is advocated for any excised GCT.

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