Atypical Granular Cell Tumor with Intranuclear Inclusions: A Case Report

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Abstract
Atypical and malignant granular cell tumors are relatively uncommon constituting 1-2% of all granular cell tumors. We present a case of 57 year old man known to have squamous cell carcinoma of anal canal in 2008, now presenting with a chest wall nodule of 2 months duration. Fine needle aspiration of the chest wall nodule yielded atypical pleomorphic cells with abundant granular cytoplasm and vesicular nuclei with prominent nucleoli and intranuclear inclusions. A biopsy was advised and tru cut biopsy proved it to be atypical granular cell tumor, positive for S100 and KP1. We present this case to increase the awareness of this uncommon entity presenting as a subcutaneous nodule.

Keywords: Atypical Granular cell tumor, Intranuclear inclusions, FNAC.

Introduction
Granular cell tumor is an uncommon tumor that was first described by Weber in 1854 and established as a clinical entity by Abrikorosoff in 1926 who termed it as granular cell myoblastoma. The common location of granular cell tumor is the oral cavity but it can also present as slowly growing solitary nodules in head and neck, chest wall and arms. They develop in breast in about 5% of cases mimicking carcinoma. Other sites are vulva, scrotum, lung etc. We present an unusual case of atypical granular cell tumor presenting as a chest wall nodule and mimicking melanoma cytologically.

Case Report
The case is that of a 57 year old man who underwent surgery for squamous cell carcinoma of anal canal in 2008 presenting now to the outpatient clinic with a chest wall nodule. The clinician was suspecting cutaneous metastasis and referred him for FNAC. It was a 3X2 cm, mobile, well defined swelling. Fine needle aspiration yielded atypical pleomorphic cells having abundant granular cytoplasm and vesicular nuclei with prominent nucleoli and intranuclear inclusions. A biopsy was advised and tru cut biopsy was done. Histopathology section showed cells with abundant granular cytoplasm and vesicular pleomorphic nuclei with intranuclear inclusions. Mitosis was less than 2/HPF and there was no necrosis. The tumor cells were focally positive for PAS stain. Immunohistochemically the cells showed diffuse cytoplasmic positivity for S100 and focal cytoplasmic positivity for KP1 ruling out the possibility of amelanotic melanoma which was the closest cytological differential diagnosis. (fig 3) MIB-1 labelling index was 10% (Fig.4). There were only 2 histological criteria namely...
pleomorphism and vesicular nuclei with intranuclear inclusions putting it under atypical granular cell tumor. The patient was lost to follow up.

Fig 1. FNA chest wall pap 400x cells showing granular cytoplasm and intranuclear inclusions

Fig 2. Trucut biopsy. H&E 200x cells have granular cytoplasm and intra nuclear inclusion

Fig 3a. Trucut biopsy S100 positive

Fig 3b. Trucut biopsy 400x KP1 positive

Fig 4. MIB 1 labelling index less than 10%

Discussion
Granular cell tumor is an uncommon schwannian benign neoplasm with predilection for upper aero digestive tract, skin and soft tissue. Atypical and malignant granular cell tumors account for less than 2% of the granular cell tumors\(^{(4,5,6)}\). These tumors primarily affect adults even though it may be found in children also\(^{(7)}\). They are common in females and often develop between the second and sixth decades of life. They may coexist with a malignant lesion. The origin of granular cell tumor is uncertain. Based on the evidence that monoclonal antibody KP1 which recognises the lysosome associated glycoprotein CD68 reacts positively with Schwannomas and granular cell tumors, it is believed that these tumors arise from schwan cells\(^{(8)}\). Granular cell tumors stain cytoplasmically for S100 protein and are closely associated with nerves. They often present near distal nerve trunks\(^{(9)}\). All these support a schwannian cell origin.

Six histologic criteria were assessed to decide whether a tumor is benign, malignant or atypical. They are necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity (more than 2/10 HPF), high N/C ratio and pleomorphism. Neoplasms that satisfy 3 or more criteria are classified as histologically malignant, those that met 1 or 2 criteria are put under atypical and those which displayed focal pleomorphism but fulfilled none of the other criteria are termed benign\(^{(4,10)}\). Our case comes under atypical granular cell tumor as it showed 2 of the above mentioned criteria which are marked pleomorphism and vesicular nuclei with nucleoli.
and intranuclear inclusions. There was no other histologic criteria to call it malignant. The literature search showed only a few case reports of atypical granular cell tumor. Unless we are aware of this entity one can be easily misled by the cytological appearance of this tumor as in our case Amelanotic melanoma was the closest cytological differential diagnosis in our case since the cells contain prominent intranuclear inclusions. But the tumor cells had strikingly granular cytoplasm and they were focally PAS positive. Besides, the positivity of the tumor cells for both S100 and KP1 ruled out melanoma. We are reporting this case because of its rarity and the presence of intranuclear inclusions which will mislead the pathologist to give a wrong diagnosis of melanoma unless one is aware of this entity.

References

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