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MAMMOTH VASCULAR ECCRINE SPIRADENOMA - VASCULAR ECCRINE SPIRADENOMA OF UNUSUALLY LARGE SIZE

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Abstract

Context: *Giant Vascular eccrine spiradenoma is a distinct unusual variant of eccrine spiradenoma characterized by extensive vascularity and hemorrhage accompanied by a large size, as compared to the usual eccrine spiradenoma. As a result, clinically and histopathologically it can be mistaken for an angiomatous or a malignant lesion. Also the published reports of giant vascular eccrine spiradenoma depicts the fact that these giant vascular eccrine spiradenoma are no more than 5 cm in size*

Case report: *A 65 year old male patient presented with a mamoth sized i.e., 10 x 6 cm mass in the inguinal region. Clinically and on gross examination a diagnosis of angiosarcoma was made. However a diagnosis of vascular eccrine spiradenoma was made.*

Conclusion: *We propose the term mammoth vascular eccrine spiradenoma for this unusual giagantic variant of vascular eccrine spiradenoma. Awareness of the entity and histopathologic examination will prevent whirlwind of appointments with oncology and enable accurate management decisions.*

Keywords: *Giant, vascular, mammoth, eccrine spiradenoma*

Introduction

Giant vascular eccrine spiradenoma (GVES) is a very rare variant of eccrine spiradenoma and differs from the usual eccrine spiradenoma (ES) by large size and increased ascularity.[1,2,3]As a result clinicians and pathologists easily misdiagnose it as angiomatous or malignant esion.[2] Usual ES measures not more than two cm in dimension. The terminology, giant vascular ES, as first described by Cotton et al in 1986 was used for ES s measuring more than 3 cm.[1] All the published reports of GVES depicts the fact that these GVES are no more than 5 cm in size. We report a case of 65 year old male presenting with a mamoth sized i.e., 10 x 6 cm mass in the inguinal region, diagnosed as vascular eccrine spiradenoma on histopathology. Hence we propose the term mammoth vascular eccrine spiradenoma (MVES) for this unusal giagantic variant of ES. Management is dependent on a good clinical Diagnosis, and an accurate histopathology evaluation of the excised specimen.

Case Presentation

65 year old male presented with swelling in the left inguinal region of 20 years duration. There was a history of increase in the size of swelling since six months. The swelling measured 10 x 6 x 4 cm and was mobile and firm in consistency. External surface showed ulceration of one cm in size. Clinically a diagnosis of angiosarcoma was made. At surgery wide local excision was done.

Gross: Specimen consisted of single skin covered globular mass measuring 8 x 5 x 4 cm. Grossly margins were not involved. Cut surface was predominantly hemorrhagic, solid with focal myxoid areas and cystic change. (Figure 1) Entire tumor was step sectioned and processed for routine histopathology.

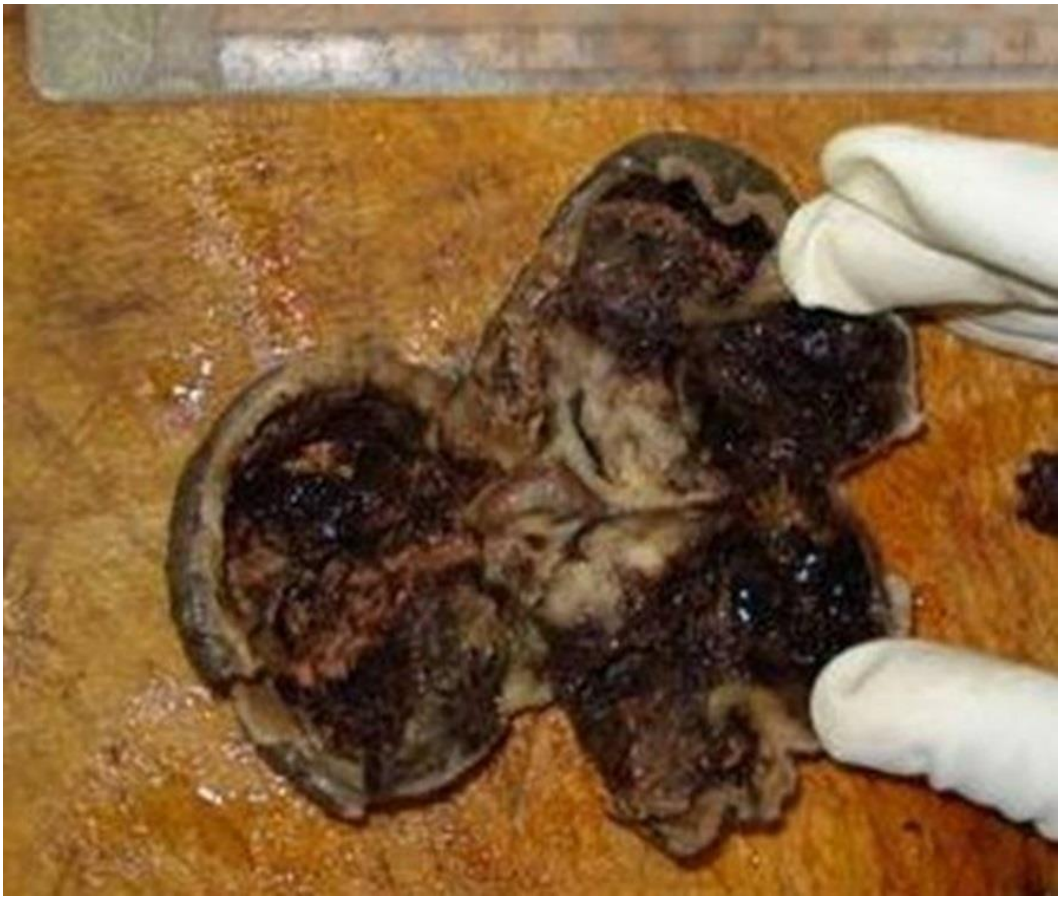

Figure 1: Gross photograph of the cut surface showing predominantly hemorrhagic, solid with focal myxoid areas and cystic change

Microscopy: Sections studied showed superficial skin which was unremarkable along with a well circumscribed, encapsulated, subepithelial tumor. (Figure 2 a) Tumor was composed of extensive areas of vascular formations along with solid areas of tumor cell proliferation.The vascular areas showed many venous caliberated blood vessels lined by flattened endothelium and filled with blood. (Figure 2b) Many of these blood vessels show anastomosing channels.The tumor islands show tumor cells arranged predominantly in solid nests and islands separated by fibromyxoid stroma.the tumor islands show peripheral dark oval cells with scanty cytoplasm and central pale cells with moderate cytoplasm and pale nuclei. (Figure 2c) Abundant thin pink basement membrane material is seen entering in between the tumor cells. (Figure 2d)No evidence of mitosis, necrosis or aytpia noted.

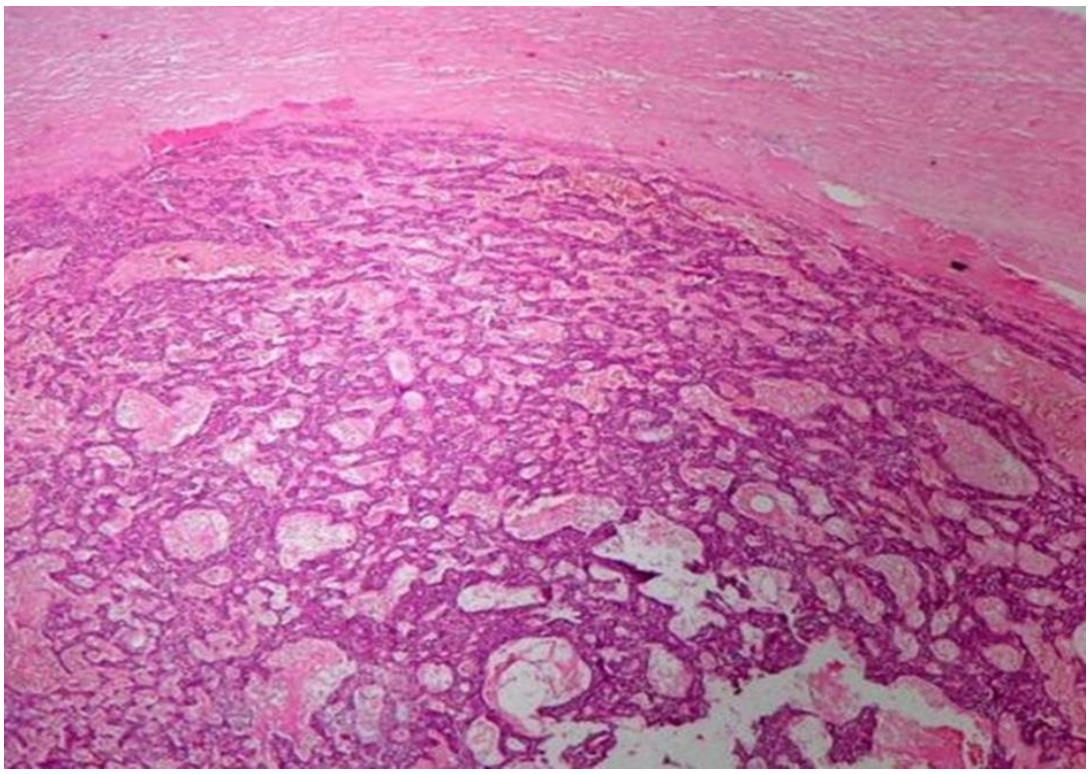

Figure 2a: Microphotograph showing well circumscribed, encapsulated, subepithelial tumor (H&E x 40).

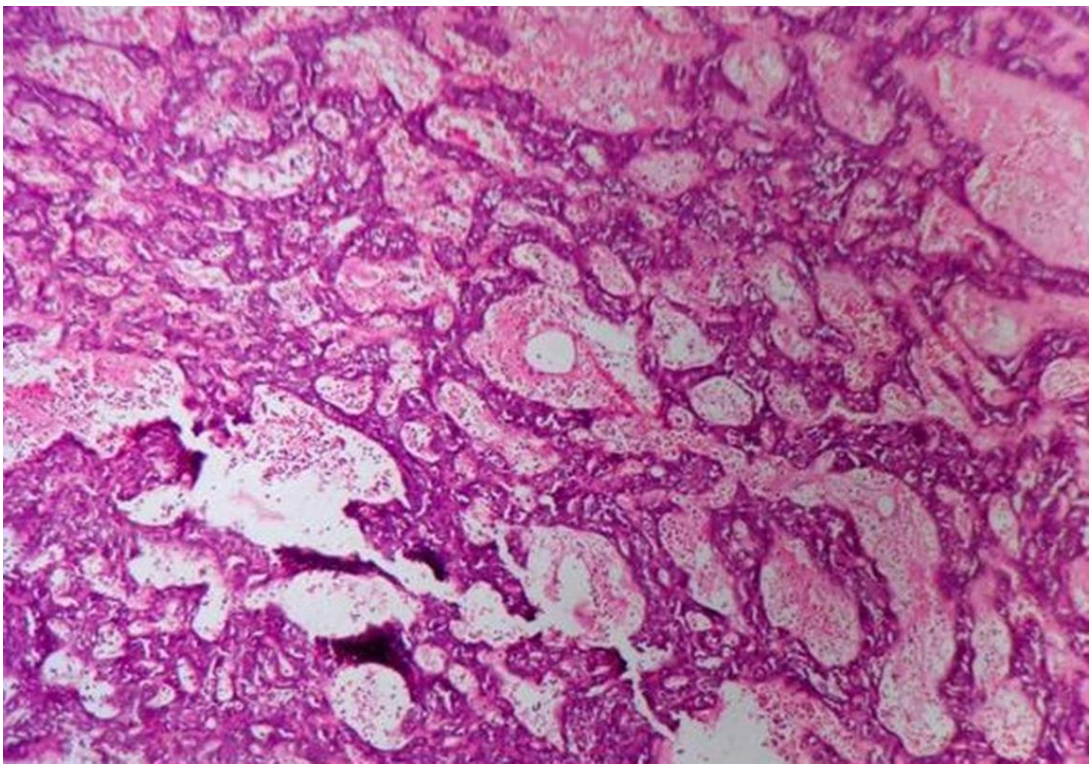

Figure 2b. Microphotograph showing many venous caliberated blood vessels lined by flattened endothelium and filled with blood. (H&E x 100).

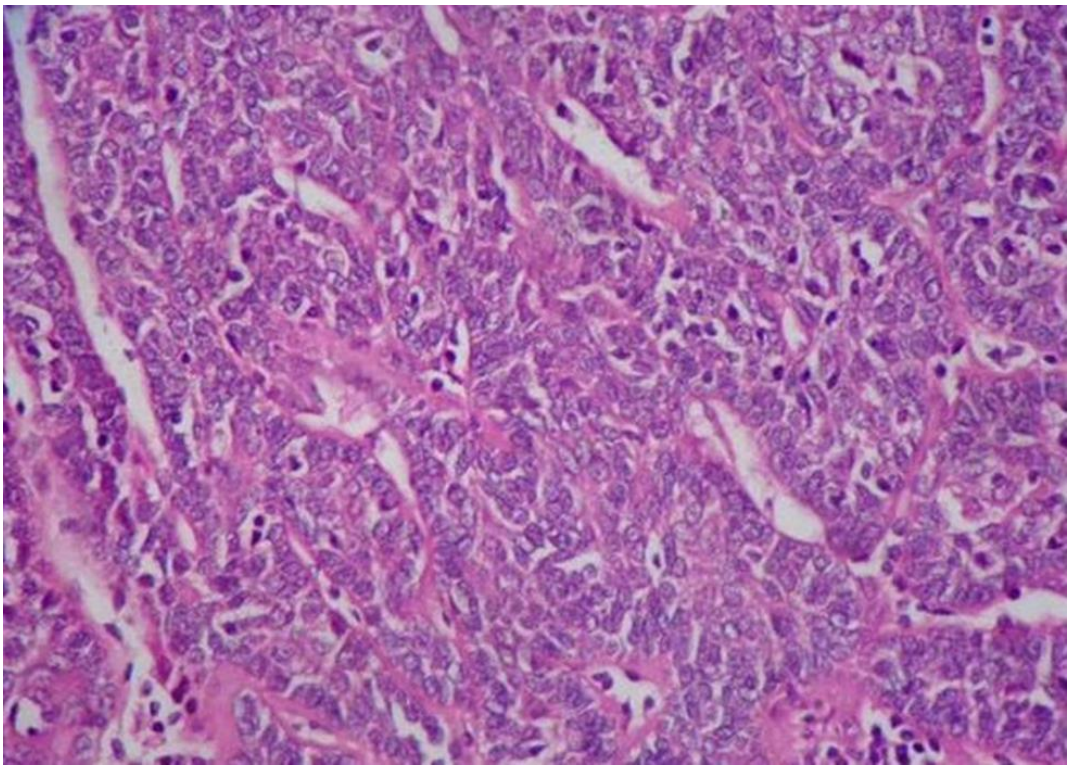

Figure 2c: Microphotograph showing tumor islands with peripheral dark oval cells with scanty cytoplasm and central pale cells with moderate cytoplasm and pale nuclei (H&E x 200).

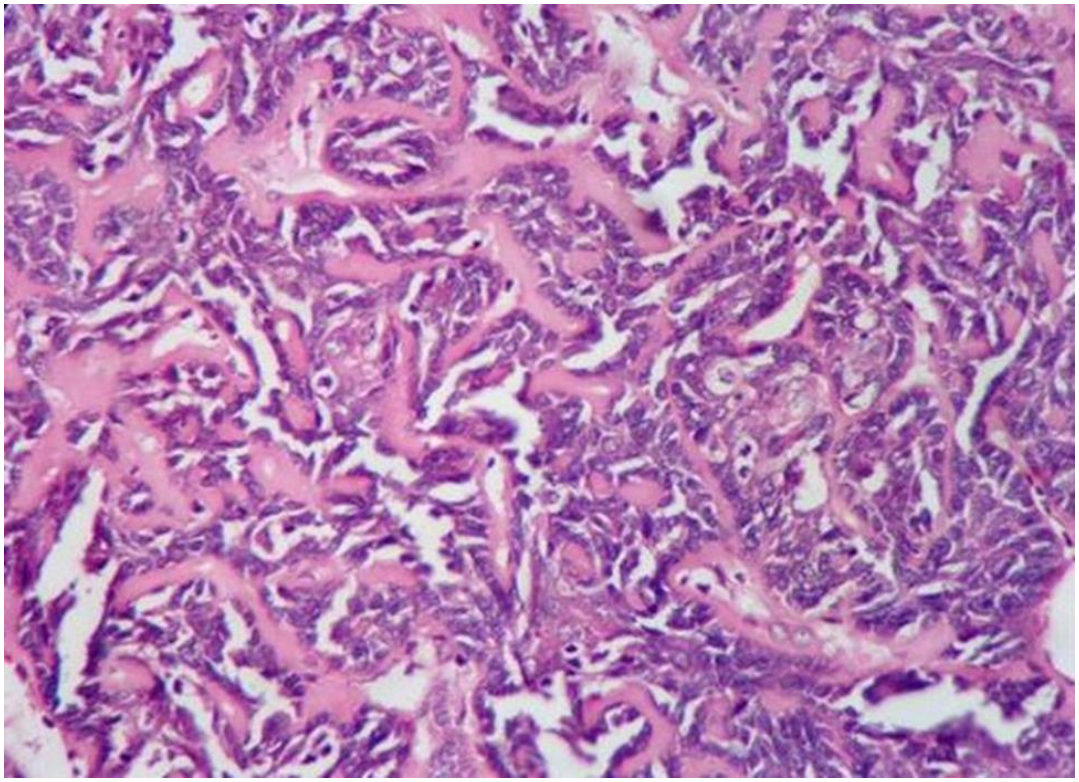

Figure 2d: Microphotograph showing abundant thin pink basement membrane material is seen entering in between the tumor cells (H&E x 200).

Hence a final diagnosis of Mammoth vascular eccrine spiradenoma was made. Patient is doing well 10 months post-surgery.

Discussion

Adnexal tumors present unique difficulties, in part related to the wide variety of tumors, the substantial frequency of one lesion exhibiting histologic pattern of two or more adnexal lesions and complicated nomenclature. GVES is an unusually rare variant of eccrine spiradenoma characterized by extensive vascularity and hemorrhage accompanied by a large size, as compared to the usual ES. There are only six reports of GVES published in English literature. [1, 2, 4, 5 and 6] The lesion is usually mistaken for an angiomatous lesion or malignancy by both clinicians and pathologist thus rendering difficulties in planning the management.

All the six reported cases of GVES occurred in adults and elderly population. GVESshas been reported to occur more frequently in the trunk and extremities followed by back of scalp and costochondral line occasionally.[3]In contrast to ES which usually presents as a painful; slow growing mass without skin changes of bleeding and ulceration,[7]GVES presents many a times as asymptomatic painless mass.[1]At times, GVES may manifest with bleeding and ulceration which misleads a clinician to make an erroneous diagnosis of malignancy.[1,6]

In all the previous reported cases, the sizeof GVES was no more than 5 cm. However in the present case report since the size of the tumor was more than 10 cm in diameter, the term mammoth VES was used.

Taking into concern these peculiar gross features of unusual large size, increased vascularity and ulceration the lesions are mistaken for an angiomatous lesions like angiosarcoma or malignant melanoma as in our case. Careful attention to the histopathology plays a key role in management decisions.

The cell of origin may apparently express one or more lines of appendegial differentiation indicating that these cells are not a priori restricted to one line of differentiation. Historically, IHC has been of little value in distinguishing among the phenotypic patterns of adnexal neoplasm. [7]

To, unravel the mystery of histogenesis, Joo Youun et al, who performed IHC using markers for epithelial, small basal cells and myoepithelial cells and concluded that GVES originates from eccrine gland and differentiates towards secretory portion of secretory coil. [2]

Taken together, the increased vascularity in GVES points to the fact that the tumor may be arising from the highly vascular portion of eccrine sweat gland as secretory part of the eccrine duct. [2]

Malignant degeneration has not been reported yet in the GVES. However, like in a usual benign ecccrine spiradenoma, adequate sampling of the entire tumor is must to prevent missing a focal malignant change in a benign lesion. [8]

Conclusion

MVES is an unusual very large variant of ES and should be considered in the differential diagnosis of any soft tissue mass which appears hemorrhagic on gross appearance. With challenging conditions and diseases to diagnose, those working as pathologist learn something new every day. Histopathological diagnosis is must for accurate diagnosis and management.

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