Massons tumor with variable morphology

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ABSTRACT

Massons tumor is a benign reactive intravascular endothelial proliferation. It is an unusual pattern of organization of a thrombus. We report a case of a 15-year-old boy with a small tumor nodule in the left temporal region of the face. Cytology was reported as fibrohistiocytic tumor. Histopathology showed the classic features of Massons tumor. In addition, there were onion whorl like areas and myxoid change. We have also highlighted the cytological features on a retrospective review.

Key words: Massons tumor, reactive intravascular endothelial proliferation, onion skinning

INTRODUCTION

Massons tumor is unusual benign reactive intravascular endothelial proliferation. Pierre Masson first described the tumor in 1923. [1] It represents 2% of the vascular tumors. [2] This tumor is usually misdiagnosed clinically and cytologicaly. The importance of identification of this tumor is to differentiate this benign proliferation from Dabskas tumor and angiosarcoma. We report a case of Massons tumor in a 15-year-old boy with an additional histopathological finding of onion whorl pattern of proliferation of the endothelial cells. We have also highlighted the presence of vascular fragments with papillae on cytology smears on a retrospective review.

CASE REPORT

A 15-year-old male presented with a left temporal region swelling for 2 months. Clinical diagnosis was a dermoid cyst. On examination, there was a 1 cm diameter swelling in the subcutaneous plane mobile nontender and soft. The overlying skin showed gray brown discoloration. There was no history of trauma.

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Cytology

Fine-needle aspiration was hemorrhagic with few vascular fragments. There were loosely cohesive and scattered scanty cells, round with eccentric nuclei resembling reactive fibroblasts/histiocytes. Diagnosis of a benign fibrohistiocytic tumor was made.

Histopathology

Gross examination of the resected nodule showed a well-circumscribed lesion, 2 cm in diameter, solid, gray white with areas of hemorrhage. Microscopic examination revealed a well-defined large vessel wall with an organizing thrombus [Figure 1]. Large areas of the thrombus had papillary endothelial hyperplasia with fibrin cores [Figure 2]. Many of the small vessels had marked endothelial proliferation and swelling with onion skinning resembling hyperplastic arteries [Figures 3 and 4a]. Foci of myxoid change were seen in the stroma [Figure 4b]. The endothelial cells were plump with prominent nucleoli in few [Figure 4c]. However, there was no necrosis, anastomotic channels and mitosis. The final diagnosis of Massons tumor was made.

Retrospective review of the cytology slides showed vascular fragments with tiny papillae in the papanicoloau stained smears [Figure 4d]. Cellular morphology was difficult to appreciate within the papillary fragments [Figure 5a]. The core was granular and covered by endothelial cells [Figure 5b]. The reactive endothelial cells were very plump with anisocytosis and prominent nucleoli in few of them [Figure 5c]. Intracytoplasmic lumina and hemosiderin granules were absent. There was also scattered basement membrane-like material [Figure 5d].

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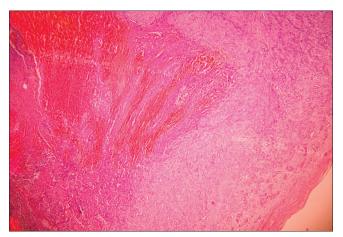


Figure 1: Section shows dilated vessel wall with thrombus. The thrombus is showing areas of the organization (H and E, ×40)

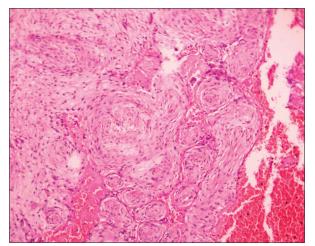


Figure 3: Section shows prominent onion whorl pattern of endothelial cells (H and E, ×100)

DISCUSSION

Massons tumor represents a florid pattern of reaction of endothelial cells in an organizing thrombus. The causative factors considered in the literature are minor trauma, [11] estrogenic influence paracrine/cytokine influence like fibroblast growth factor. [2] It can be primary or occur on preexisting vascular lesions. In our case, there was no history of trauma or preexisting vascular lesion.

Hashimoto et al., have classified it into three types.[2]

Type 1 - Primary form occurring within dilated vessel

Type 2 - Secondary form occurring on preexisting vascular channels

Type 3 - Extravascular location secondary to trauma/hematoma

Our case was Type 1 the most common type.

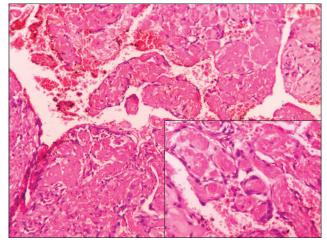


Figure 2: Section shows areas of papillary endothelial hyperplasia. Inset shows the fibrin core of the papillae lined by plump endothelial cells (H and E, ×100, ×400)

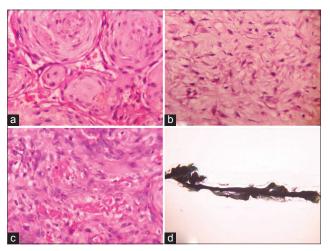


Figure 4: (a) Section shows high power view of the onion whorl pattern of endothelial cells. (b) Foci of myxoid change. (c) Plump endothelial cells with prominent nucleoli (H and E, ×400). (d) Vascular fragment with tiny papillae in cytology smear (PAP, ×40)

Clinical features

The tumor can occur at any age and has equal incidence in males and females. The average age group of incidence is 42 years. The common sites of occurrence are head, neck, and extremities. It usually presents as a firm tender nodule with red to brown discoloration of the overlying skin. Our patient was a young boy with lesion in the head and overlying skin discoloration of the nodule was present.

Cytology

Cytology smears are usually hemorrhagic, and many show few vascular strands and fibrin.^[3] Our case in addition had clusters and scattered plump cells resembling reactive fibroblasts/fibrohistiocytic cells which mislead the diagnosis. On review, they represented the reactive endothelial cells. The presence of papillary structures attached to the vascular fragments has not been previously described in the literature to the best of our knowledge.

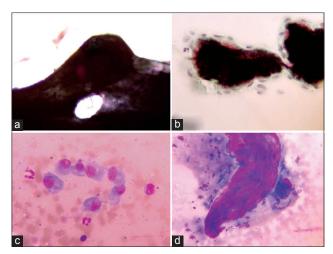


Figure 5: (a) Section shows papillary fragments in cytology smear with a granular core. (b) High power view of the papillae lined by endothelial cells (PAP, ×400). (c) Clusters of plump endothelial cells in a hemorrhagic background. (d) Basement membrane material (MGG, ×400)

Histopathology

Microscopically it shows endothelial proliferation forming papillary tufts. The core of the papillae is made up of fibrin which is a characteristic feature. Our case had a similar appearance, and in addition had a prominent onion whorl appearance of plump endothelial cells similar to hyperplastic arteriolitis. This kind of reaction has not been previously described in the literature to the best of our knowledge.

The importance of Masson's tumor is to differentiate it from Dabska's tumor and low-grade angiosarcoma. Massons tumor is well-circumscribed with plump endothelial proliferation. There is no atypia, necrosis, and anastomosing channels as seen in angiosarcoma. [2,4]

CONCLUSION

The florid proliferation of the endothelial cells in Massons tumor can sometimes form onion whorl like appearance. Careful observation of vascular fragments with papillae on cytology smears in a hemorrhagic background, basement membrane material, reactive endothelial cells, and correlation with clinical findings can suggest the diagnosis on cytology.

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