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CASE REPORT

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Calcifying fibrous pseudotumor in association with hyaline vascular type *Castleman's disease*

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Abstract

Calcifying fibrous pseudotumor is a recently described rare entity. It is considered as sclerosing end stage of inflammatory myofibroblastic tumor. An association with hyaline vascular type Castleman's disease has also been described. We report a case of a 13-year-old boy who presented with pain in epigastrium. Computed tomography scan of abdomen revealed a circumscribed mass arising from the gastric wall along the greater curvature. Histology revealed a tumor composed of spindle cells present within the dense hyalinized collagenous tissue. Lymphoplasmacytic infiltrate was seen along with lymphoid follicles, dystrophic and ossifying calcification. Tumor cells were focally positive for alpha smooth muscle actin and negative for anaplastic lymphoma kinase protein.

Keywords: Calcifying, Castleman's disease, pseudotumor

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Introduction

Calcifying fibrous pseudotumor is a rare, recently described benign tumor. [1] Reports have shown association with hyaline vascular type Castleman's disease. [2] It usually occurs in children and young adults. We report a case of calcifying fibrous pseudotumor presenting in association with hyaline vascular type Castleman's disease.

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Case Report

A 13-year-old boy presented with history of retro-sternal burning sensation, postprandial epigastric pain, weakness and pallor of two years duration. Symptoms gradually became worse. A mass in the epigastrium was also noted. X-ray chest showed a calcified mass in the left hypochondrium. Unenhanced computed tomography scan of abdomen revealed a circumscribed mass arising from the gastric wall along the greater curvature in the region of fundus [Figure 1]a. The lesion appeared to be densely calcified. Lymph nodes were seen in the splenic hilum.

The mass was excised with a healthy rim of gastric wall. Splenectomy was also performed. On gross examination, a well-circumscribed, 8.0 x 6.0 x 6.0 cm tumor was seen arising from the gastric wall with ulceration of the overlying mucosa. The tumor was adherent to the spleen, pushing the splenic parenchyma. However, there was no infiltration of the spleen seen on gross examination. [Figure 1]b Cut surface of the tumor was firm and tan gray with extensive central calcification leaving a rim of soft tissue around. Cut surface of spleen appeared unremarkable. Enlarged lymph nodes were seen in the splenic hilum; the largest measured 2.0 x 2.0 x 1.5 cm.

Histological examination revealed a circumscribed, partially encapsulated, paucicellular neoplasm characterized by dense eosinophilic hyalinized collagenous tissue with sparse spindle cells [Figure 2]a. Extensive dystrophic and ossifying calcifications were noted [Figure 2]b. Periphery of the tumor and the areas beneath the gastric mucosa revealed more cellular bundles of cytologically bland spindle cells present in a vague fascicular arrangement. The cells had oval to elongated nuclei and amphophilic cytoplasm. There was dense lymphoplasmacytic infiltrate with scattered lymphoid follicles with germinal centers. Some follicles showed hyalinization of germinal centers with concentric onion skin arrangement of mantle zone lymphocytes. The tumor was adherent to and pushing the splenic parenchyma; however, a thick capsule was seen separating the tumor from the spleen. Immunohistochemistry was performed. The tumor cells showed negativity for anaplastic lymphoma kinase protein and cytokeratin. Focal reactivity for alpha smooth muscle actin was seen [Figure 2]c.

The submitted lymph nodes on histological examination showed altered architecture by an increased number of lymphoid follicles having small germinal centers containing hyaline material with paucity of lymphoid cells and concentrically arranged mantle zone cells. A few germinal centers were radially penetrated by hyalinized blood vessels (lollipop follicles) [Figure 2]d. Interfollicular areas showed increased number of venules with scattered plasma cells. On histological examination, the spleen was unremarkable. Based on histology and immunohistochemistry, the case was diagnosed as calcifying fibrous pseudotumor (CFP).

On follow-up, the patient recovered completely, had gained weight and is asymptomatic after 12 months.

Discussion

Calcifying fibrous pseudotumor (CFP) is a rare benign fibrous lesion. The tumor usually affects children and young adults. The size in various published reports ranges from 2.0 to 15.0 cm. Simple excision is the treatment of choice. Occasional recurrences are recorded. This tumor was first described by Rosenthal and Abdul-Karim [3] under the term "Childhood fibrous tumor with psammoma bodies" in 1988. Fetsch *et al.* [1] renamed the same as calcifying fibrous pseudotumor in 1993. The pathogenesis remains unknown. However, the underlying process is likely to be fibro-inflammatory and reactive in nature. There are examples which have followed trauma. [4] A possible relationship with other pseudotumors like nodular fasciitis and inflammatory myofibroblastic tumor has been proposed. It has also been suggested that this lesion may represent sclerosing end stage of inflammatory myofibroblastic tumor. Multiple peritoneal CFPs and inflammatory myofibroblastic tumors were described in a 17-year-old girl by Van Dorpe *et al.* [5] Transitional stages between CFP and inflammatory myofibroblastic tumor were also noted within the same lesion. This case illustrated a histogenetic relationship between CFP and inflammatory myofibroblastic tumor. Pomplun *et al.* [6] also showed features of both tumors within the same lesion. Our case showed hyalinized fibrocollagenous and paucicellular areas with extensive dystrophic calcification as well as more cellular areas at the periphery. Presence of cellular and paucicellular areas raises the possibility of transition from inflammatory myofibroblastic tumor to CFP.

Association of CFP with hyaline vascular type Castleman's disease has also been recorded, illustrating the reactive nature of both lesions. [2] Kocova *et al.* [7] reported a case of CFP of visceral peritoneum focally showing lymphoid tissue, featuring changes suggestive of hyaline vascular type Castleman's disease. A case of CFP arising in a lymph node involved by hyaline vascular type Castleman's disease has been reported by Dargent *et al.* [2] In our case, the lymph nodes showed hyaline vascular type Castleman's disease and the tumor had lymphoid follicles with a few hyalinized germinal centers mimicking hyaline vascular type Castleman's disease. These changes again complement the possibility of a probable relationship between the two entities.

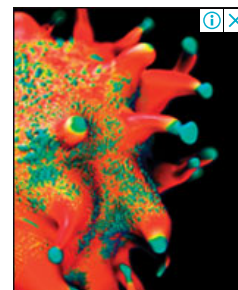
Considering all these findings, our case is unique in respect to the presence of calcifying fibrous tumor with areas suggestive of transition from inflammatory myofibroblastic tumor and associated hyaline vascular type Castleman's disease in the lymph nodes.

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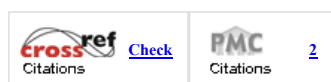
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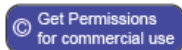
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Figures

[\[Figure 1\]](#), [\[Figure 2\]](#)

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