



Rare Case of Inflammatory Myofibroblastic Tumor of the Adrenal Mimicking Adrenocortical Carcinoma

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Abstract

Background: Inflammatory Myofibroblastic tumors (IMT) arising from the adrenal gland are extremely rare. Until now only 5 cases have been reported in literature.

Case Report: 55 year old male under evaluation for fever, right upper abdominal pain and non-progressive jaundice on abdominal CECT was found to have a large right suprarenal mass, high suspicion of adrenocortical carcinoma. He underwent adrenalectomy after hormonal evaluation. Final histopathology was Inflammatory Myofibroblastic tumor. A rare tumor composed of plump, spindle cells or myofibroblasts and histiocytoid cells arranged haphazardly and in fascicles, accompanied by prominent chronic inflammatory infiltrate, particularly plasma cells.

Conclusion: IMT of the adrenal are rare. Radiologically and clinically they mimic a malignant process and must be considered in the differential of radiologically suspicious adrenal masses. Complete surgical resection is mandatory as malignancy cannot be excluded preoperatively.

Keywords: Inflammatory myofibroblastic tumors; Adrenal gland; Histopathology; Immunohistochemistry

Background

Inflammatory Myofibroblastic tumors (IMT) are an uncommon spindle-cell proliferation that may involve any site - abdomen, lungs, orbit, gastrointestinal and genitourinary tracts [1]. Usually seen in children and young adults. The advent of immunohistochemistry brought increasing awareness of its distinctive features [2]. The adrenal gland is a rare primary site, with only four reports in the world literature [1,3-5]. Clinical differential diagnoses include adrenocortical tumours, pheochromocytoma, adrenal metastases and tuberculosis. Imaging features are generally nonspecific, preoperative differentiation from other adrenal masses is very difficult. Complete surgical resection is therefore mandatory as malignancy cannot be excluded preoperatively. We report a fifth case of this rare entity.

Case Presentation

55 year old, male, from a small town in India was being evaluated for fever, right upper abdominal pain and jaundice of 14 days duration. He underwent an Ultrasonography of abdomen which showed large liver SOL and was referred to the Gastroenterology clinic at our Institute. Fever was high grade, intermittent type, not associated with chills and rigor. Upper abdominal pain was acute onset, right sided, continuous dull aching in nature. No radiation, shifting or referred pain. Jaundice was non progressive, associated with loss of appetite, nausea and malaise. Not associated with clay colored stools, pruritis or abdominal distension. No history of loss of weight, hematemesis or melena. No history of similar illness in the past. No history of Tuberculosis or any other chronic illness. He was recently diagnosed with Hypertension. No history of smoking or alcohol. General physical examination was unremarkable except for icterus. Abdominal examination revealed hepatomegaly. Lower border of liver was palpable 5 cm below the right costal margin. No other organomegaly or mass abdomen palpable. Blood work up showed raised total and direct bilirubin with raised liver enzymes and normal coagulation parameters and viral serology. Patient was managed on the lines of acute viral hepatitis and liver functions returned to normal levels over two weeks with supportive treatment. To characterise the, a CECT Abdomen (Figure 1) was done which showed 17.5 cm × 16.6 cm × 13.5 cm heterogeneous peripherally enhancing lesion with multiple septations possibly arising from right adrenal with subcentimetric abdominal and mesenteric lymphadenopathy and patient

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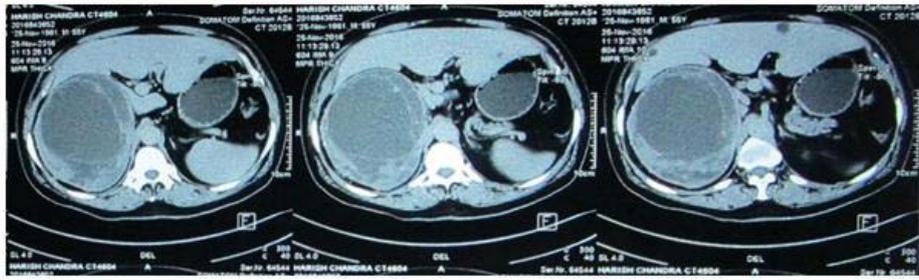


Figure 1: Heterogenous (17.5 cm x 16.6 cm x 13.5 cm) peripherally enhancing lesion with multiple septations arising from right adrenal with subcentimetric abdominal and mesenteric lymphadenopathy. Lesion closely abutting upper pole of right kidney with no clear plane from right lobe of liver. High suspicion of adrenocortical carcinoma. Opposite adrenal normal.

was referred to us. Hormonal evaluation including ONDST, serum DHEA and urinary metanephrin and nor metanephrin were normal. A provisional diagnosis of ACC was made and patient was taken up for surgery. He underwent exploratory laparotomy via Thoraco-abdominal approach. No ascites or peritoneal or mesenteric nodules or lymphadenopathy. Liver surface had multiple 1x1 cm firm nodular lesions in both lobes? hemangiomas. Tumor mass was seen arising from right adrenal with multiple parasitic vessels, closely abutting the IVC and pushing the right kidney inferiorly. Tumor was easily separable from renal vessels and kidney. No invasion/ adhesions with surrounding structures proceeded to right adrenalectomy. No RPLND was done. Grossly (Figure 2) the tumor was well encapsulated. Measuring 20 cm x 18 cm x 16 cm mass with variegated consistency, weighing about 2 kg. Cut section showed heterogenous mass with mixed solid cystic areas with areas of hemorrhage and necrosis. Pale areas interspersed in between. Intraoperative and immediate postoperative period was uneventful. Patient was discharged on sixth postoperative day.

Final histopathology was inflammatory myofibroblastic tumor, showing vague fascicles of benign spindle cells displaying plump oval to elongated nuclei, dispersed chromatin and moderate amount of cytoplasm associated with a moderate lymphoplasmacytic inflammatory cell infiltrate, with focal areas of haemorrhage and inconspicuous mitosis. Cystic areas lined by palisading fibroblasts (Figure 3). Part of normal adrenal gland identified in the periphery. Single lymph node identified was suggestive of reactive lymphoid hyperplasia. IHC (Figure 4) was positive for Desmin, ALK-1 and focal positivity for smooth muscle actin (SMA). CD 34 negative, Ki-67 proliferation index was 3-4%.

Discussion

In 1954, Umiker and Iverson coined the term “inflammatory pseudotumor” because the clinical and imaging findings mimicked those of malignant tumors [6]. IPT has been described by various names in the literature: plasma cell granuloma (heart and lung), inflammatory myofibroblastic tumor (lung), inflammatory myofibrohistiocytic proliferation, histiocytoma, xanthoma, fibroxanthoma, fibrous xanthoma, xanthogranuloma, xanthomatous pseudotumor, plasma cell-histiocytoma complex (lung), plasmocytoma, solitary mast cell granulomas, and inflammatory fibrosarcoma (urinary bladder) [7]. The exact cause of IPT is unknown. The confusion in nomenclature is a result of ambiguity in determining the process of origin of these tumors. There are two schools of thought: Post inflammatory reactive process vs. true neoplasms. Post inflammatory theory is supported by the fact that tumors are seen to occur post surgery, trauma and

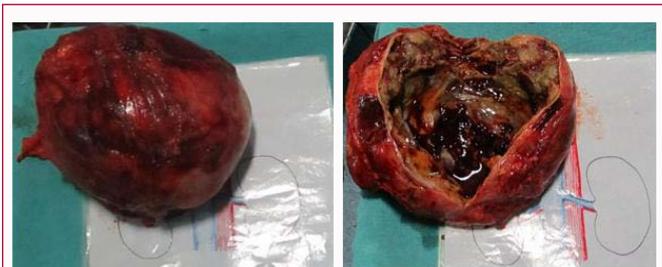


Figure 2: (A) Grossly the tumor was well encapsulated. Measuring 20 cm x 18 cm x 16 cm mass, variegated in consistency, weighing about 2 kg. (B) Cut section showed heterogenous mass with mixed solid cystic areas with areas of hemorrhage and necrosis. Pale areas interspersed in between.

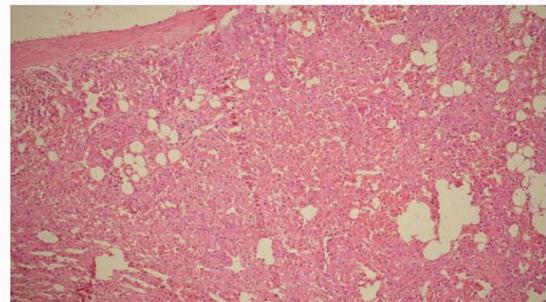


Figure 3: Histopathology showed (H&E stain, 10x) vague fascicles of benign spindle cells with plump oval to elongated nuclei, dispersed chromatin, moderate cytoplasm and lymphoplasmacytic inflammatory infiltrate with focal areas of haemorrhage and inconspicuous mitosis. Part of normal adrenal gland identified in the periphery.

VP shunts. Various organisms have been implicated in pathologic specimens, including mycoplasma and nocardiae in lung pseudotumors, actinomycetes in liver, Epstein-Barr virus in splenic and nodal pseudotumors, and mycobacteria in spindle cell tumors. IL-1 contributes to the local and systemic effects. On the other hand it has potential for local recurrence and distant metastases supporting the neoplastic theory. Most definitive evidence of neoplastic origin rests on the frequently seen clonal alterations involving the anaplastic lymphoma kinase (ALK) gene located at chromosome 2p23 in a significant subset of cases [8]. The term ‘inflammatory myofibroblastic tumour’ (IMT) was introduced in 1986 to describe lesions previously known as plasma cell granuloma or inflammatory pseudotumour [9]. IMTs are uncommon tumours of spindle-cell proliferation with predominance of myofibroblasts and histiocytes that may involve any site abdomen, lungs, orbit, gastrointestinal and genitourinary tracts. Most commonly seen in the lungs of children and adolescents, but it can occur in older persons [2,7].



Figure 4: Immunohistochemistry strongly positive for (A) Desmin, (B) ALK-1 and (C) focal positivity for SMA.

IMTs are usually asymptomatic, although 10% to 20% of cases are associated with pyrexia and weight loss. Tumour size at diagnosis is most often in the range of 5 cm to 10 cm. The biological behavior is indeterminate. Clinical differentials include adrenocortical tumours, pheochromocytoma, adrenal metastases and tuberculosis [1].

Imaging features are generally nonspecific and preoperative differentiation from other adrenal masses is very difficult. The radiologic features of IMT are variable and nonspecific possibly because of the amount of fibrosis and cellular infiltration. On ultrasound images, lesions can be hypoechoic or hyperechoic with ill-defined or well-circumscribed borders with increased vascularity during Color Doppler examinations. CECT shows varying appearances with lesions showing low, equal, or high attenuation compared with the surrounding tissues. Calcification can occur and is more common in children. Post contrast images show a variety of patterns with early peripheral and delayed central filling or heterogenous, homogeneous and no enhancement at all. Larger lesions may have central necrosis [10]. On MRI, IMTs usually have low signal intensity on both T1- and T2-weighted images, which may reflect the fibrotic nature of these lesions [11]. IPT are FDG-avid on PET/CT images. PET/CT is highly sensitive but has low specificity for IPT [12]. Complete surgical resection is mandatory as malignancy cannot be excluded preoperatively [1]. The diagnosis of IMT is almost always a histological surprise, as seen in our case. Surgical resection is recommended even in cases with recurrence [11]. There is no role of Radiation or Chemotherapy described for adrenal tumours. Radiation and corticosteroids have been used to treat patients of Lung IMTs who cannot undergo surgery. However there is no proven survival benefit compared with surgery. With radical excision, the risk of relapse is low, but relapse and distal metastasis occur many years after treatment [12].

Microscopy shows circumscribed tumour composed of plump, spindle cells or myofibroblasts and histiocytoid cells arranged haphazardly or in fascicles with intervening thick collagen bundles, accompanied by prominent infiltrate of chronic inflammatory cells, particularly plasma cells (best distinguishes IMT from fibromatosis and fasciitis). The advent of immunohistochemistry brought increasing awareness of its distinctive features. IMTs usually express actin and also desmin and keratin. Our case was positive for desmin, ALK and SMA. ALK gene rearrangements, resulting in over expression of ALK protein, are present in 30% to 40% of cases, mainly seen in pediatric

age group. IMTs that have metastasized are most often ALK negative [2,15] Hence ALK positivity can be considered a good prognostic factor. No reported recurrences for adrenal IMTs [1]. Other sites, 10% to 25% of patients have local recurrence. Less than 5% of cases metastasize, but their behavior is difficult to predict on morphological grounds, and it is increasingly believed that all cases of IMT are best regarded as low-grade sarcomas. Sarcomatous degeneration has been reported in IMTs at other sites, hence surveillance is crucial.

Conclusion

IMTs of the adrenal are rare. Clinically and radiologically they mimic a malignant process and must be considered in the differential of radiologically suspicious adrenal masses. Complete surgical resection is mandatory as malignancy cannot be excluded preoperatively. Recurrence or Metastasis of adrenal IMTs is unknown but in view of metastatic potential of lesions at other sites, careful postoperative follow up including repeat imaging at 6 months to exclude local recurrence is advised.

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