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INTRODUCTION

Amyloidosis describes a group of disorders characterized by the extra cellular deposition of amyloid substance which results in tissue damage. It has been classified as acquired systemic, organ limited or localized.[1]

The localized form of amyloid deposit as a mass/nodule in the absence of systemic involvement is known as amyloidoma.[2] Amyloidoma has been described in multiple body sites including respiratory, genitourinary, gastrointestinal tract, central nervous system, skin and breast. Amyloidoma of soft tissues is exceedingly rare and occurs mostly in the mediastinum and retroperitoneum.[3] There has been no previously published case reports of abdominal wall involvement thereby we are describing a case of primary localized amyloidoma which presented as an abdominal wall abscess without any evidence of systemic disease.

CASE REPORT

The patient was 20-year-old male with a history of painful rounded abdominal mass since 2 years which had begun growing since last 6 months. The patient had moderate splenomegaly. The clinical signs and CT scan strongly suggested soft tissue abscess, likely to be caused by cysticercosis or a neoplastic process. The mass was excised and sent for histopathology. Grossly we received tan-colored nodular mass measuring 6.5 × 5 × 3 cm. Cut surface showed a cavity filled with white yellow waxy material [Figure 1].

Microscopically, there was the presence of amorphous fibrillar eosinophilic material deposited in between the muscle bundles. These deposits were accompanied by massive inflammatory infiltrate rich in macrophages, lymphocytes, plasma cells and giant cells [Figure 2]. There was no evidence of immaturity and cellular atypia in the plasma cells. The fibrillar material was PAS positive and congophilic [Figure 3]. Potassium permanganate incubation caused loss of reactivity with Congo red indicating AA type amyloid deposits. The deposits gave lava red color on fluorescent microscopy [Figure 4]. Immunohistochemistry to determine the specific protein responsible for forming amyloidoma were not performed. There was no evidence of systemic amyloidosis or any underlying inflammatory condition or a neoplastic process. Further investigations including serum protein electrophoresis, Bence-Jones proteinuria were normal. Real time PCR (Q-PCR) for demonstration of Mycobacterium tuberculosis was performed and showed negative results [Figure 5]. In the 6 months since excision, the mass was not recurred.

DISCUSSION

Amyloidoma is an uncommon manifestation of amyloidosis. Abnormal glycoprotein, usually light
chain immunoglobulin, rarely serum amyloid A protein is deposited locally, forming a solitary mass. The etiology is not well defined. Some believe that the precursor protein in localized cases is made by plasma cells which are adjacent or in the deposits even if they are few in number. On the other hand, in systemic cases the precursor protein exists in the circulation and their site of production may be far from the place in which they are deposited. It is usually seen as solitary or multiple nodules in variety of anatomical sites including gastrointestinal, respiratory, genitourinary tracts. These nodules are usually large and present with signs and symptoms of pressure which compress the adjacent organs. Amyloidoma in isolation without concomitant plasmacytoma or other disease is extremely rare in soft tissues and only few cases have been reported. The largest reported series of soft tissue amyloidoma by Krishnan et al. is a study of 14 cases. Of these, seven were in the mediastinum, three in the mesentery, three in the retroperitoneum, and one in the right flank. On the basis of the associated morphologically atypical and phenotypically monoclonal cell population, the resistance to potassium permanganate pretreatment, and the lack of reactivity with anti-AA antisera, 10 cases were classified as immunocytic dyscrasia associated with AL-amyloidosis. However, only four cases had features of AA-amyloidosis. This distinction was made because patients with AA-amyloidoma of soft tissues appear to have better...
prognosis. Ghiasi et al. had reported a case of primary isolated retroperitoneal amyloidoma with osseous metaplasia without any concomitant plasmacytoma or other disease.

Our case presented as abdominal wall abscess. The clinical and radiological investigations suggested the case to be cysticercosis or some neoplastic process. Clinical suspicion of amyloidosis as a cause of abdominal abscess is very rare, thereby leading to misdiagnosis of neoplasm/cysticercosis. The present case was a diagnostic challenge because the dominant feature of the lesion was extensive acute and chronic inflammatory infiltrate and giant cell reaction. These cells overshadowed the amyloid deposits, leading to diagnostic difficulties.

Microscopically, there is no difference between the amyloid tumors present in primary or secondary amyloidosis. IHC stains may be helpful to identify the specific type of protein fibril involved in deposit. Staining is done to see the immunoreactivity of deposits to antibody against amyloid precursor protein (APP), kappa and lambda immunoglobulin light chain. In our case, microscopic finding and fluorescent study were diagnostic. Results with potassium permanganate were typical for AA amyloid.

It is important to make a distinction between AL and AA amyloidoma as patient with AL amyloidoma have poor prognosis. Krishnan et al. found that 10 of his 14 patients had AL amyloid and 80% of them subsequently developed lymphoplasmacytic malignant neoplasm. Progression to disseminated disease is also common and additional tumors develop after an interval of 3-9 months. In contrast, AA amyloidoma do not recur or develop immunocytic dyscrasia and are usually cured by excision. Management requires a complete excision of the tumor. Radiotherapy can be used to decrease the size of bone-occupying lesions. Aggressive chemotherapy has been used in cases with underlying lymphoproliferative disorders.

Although it is not a true neoplastic process, amyloidoma can have devastating consequence for the patient. Accurate diagnosis is possible with a systematic approach incorporating clinical, radiological and histopathological assessment of involved tissue. Congo red staining, fluorescent microscopy and observation of apple-green birefringence under polarized light is essential to confirm the amyloid deposits.

REFERENCES


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