Rare disease

Primary amyloidoma of chest wall – a rare condition

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Summary

Organ specific amyloidosis has been shown to be confined to various organs like liver, lung, spleen, urinary bladder and gastro-intestinal tract, but amyloidoma or tumoural amyloidosis of chest wall with no evidence of systemic amyloidosis is an extremely rare presentation and only two cases have been reported till now. The authors report a case of chest wall amyloidoma with review of literature.

BACKGROUND

Primary chest wall tumour accounts for 1–2% of the total primary tumours and approximately 60% of these masses are malignant.¹ Malignant chest wall tumours mostly arise from the soft tissue and 85% of bony chest wall tumours originate from ribs and cartilage, chondrosarcoma being the most common. Primary amyloidoma or tumour amyloidosis of chest wall is an extremely rare condition which causes local tissue destruction and can restrict chest wall expansion.

CASE PRESENTATION

A 65-year-old female was referred to the department of General Surgery, CSM (King George's) Medical University, Lucknow, India in October of 2008 for an unsuccessful excision of a progressively increasing lump in lower left chest wall from 1 year. There was no history of trauma, fever, weight loss, dyspnoea, haemoptysis and bladder-bowel complains. Her medical, social and family history was non-significant. Clinical examination revealed a mildly tender, hard lump of size 11×9×6 cm in the left lower lateral chest wall. The margins of the lump were relatively well demarcated and the surface was smooth. The lump appeared to be fixed to the underlying ribs. There was no evidence of regional lymphadenopathy.

INVESTIGATIONS

Plain skiagram chest revealed a mass at the left lower chest wall. CT scan showed an oval lump of 12×8×5 cm tumour extending from the lateral angle of the fifth to eighth rib. Fine needle aspiration and tru-cut biopsies of the chest wall mass only showed some non-representative necrotic debris, blood and histiocytes. Tuberculin skin testing with purified protein derivative injected intradermally showed a type II reaction at 72 h. Due to the diagnostic challenge, a surgical resection was necessary.

Histo-pathological examination proved the presence of amyloid deposits after staining with Congo red (figure 1) and there was no evidence of direct rib invasion by the mass. Rectal mucosa biopsy excluded the possibility of systemic amyloidosis.

DIFFERENTIAL DIAGNOSIS

Chest wall tumour (chondroma, fibrous dysplasia, osteoclastoma, osteosarcoma chondrosarcoma).

TREATMENT

A postero-lateral thoracotomy was carried out involving the old scar. A tumour involving the lateral and posterior chest wall of ribs one through five, without parenchymal involvement, was identified and resected (figure 2). The chest wall was closed with no need for reconstruction because of adequate coverage from the intercostal muscles; however reinforcement of chest wall was achieved by putting prolene mesh.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful and the patient was discharged in satisfactory general condition on tenth postoperative day and until now no local recurrence has appeared.

DISCUSSION

Primary chest wall tumours account for 1% to 2% of all primary tumours occurring in the body. Approximately 60% of chest wall tumours are malignant and about 50% of these arise from soft tissue rather than bone or cartilage.² Primary chest wall tumours generally manifest as slowly enlarging masses. Initially, most tumours are asymptomatic, although, with increasing growth, invasion of surrounding structures,

Figure 1 Congo red staining showing amyloid deposits.

Figure 2 Resected and cut open tumour mass seen filled with amyloid.

or the development of pathologic fractures, nearly all malignant tumours and 25% of benign masses are likely to become painful.¹ Chondromas, fibrous dysplasia etc are some of the common benign chest wall tumours. Amyloidosis of chest wall is an extremely rare condition. Amyloidosis is a slowly progressive disease which can lead to significant morbidity and death. Amyloid deposits are extracellular and not metabolised or cleared by the body.³ Thus, the progressive deposits eventually impair the function of the organs where they accumulate. Amyloid is derived from precursor proteins that either have an abnormal structure or are abnormally increased in the serum. About 18 different amyloid precursor proteins have been described and can also arise secondary to long-term haemodialysis, myeloma and chronic inflammatory conditions such as osteomyelitis, rheumatoid arthritis and tuberculosis.⁴

The important differential diagnoses of amyloidoma include various chest wall tumours and are difficult to differentiate them clinically and radiologically. This makes a histopathological examination necessary by congo red staining. Complete surgical excision with stabilisation and reconstruction of the chest wall as required should be

attempted, as this lesion responds best to local control and has a negligible local recurrence rate.⁵

To the best of our knowledge, only two cases of primary chest wall amyloidoma have been reported so far. Because of rarity of cases the exact aetiopathogenesis is still not known and as it is associated with systemic lymphoproliferative disorders and plasma cell dyscrasias, primarily multiple myeloma and lymphoplasmacytic lymphoma these conditions should be ruled out.⁶

Learning points

- ▶ Primary amyloidoma of chest wall is a rare disease.
- ▶ Rectal mucosa and submucosa biopsy should be done in all patients with primary amyloidoma to rule out systemic amyloidosis.
- ▶ An intraoperative biopsy technique would have helped us in diagnosing the nature of disease preoperatively and might have prevented the resection of non-involved ribs.
- ▶ All patients should also be investigated for systemic lymphoproliferative disorders.

Competing interests None.

Patient consent Obtained.

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