ABSTRACT • We report the case of a fifty-two years old patient who presented symptoms of spinal cord compression with incomplete paraplegia in relation to a solitary vertebral tumor of T5. The patient required an urgent surgery for decompression and fixation; the resection of the tumor was irradialional and was complemented by radiotherapy later on. Results of the histological examination confirmed the tumoral nature of the lesion and were compatible with the diagnosis of solitary amyloidoma of T5. The patient recovered progressively.

The disease recurred few months later with extension to the lower level T6. The patient underwent a second surgery as complete as possible because the global prognosis of this kind of tumor is essentially related to local recurrence. So a double level vertebrectomy by posterior approach only was performed.

At 2 years follow-up after the second surgery, the patient did not show any local recurrence and his neurological status was stable.

The aim of this paper is to describe a rare case of thoracic solitary amyloidoma. Diagnosis of such a tumor is not easy due to its aspecific clinical and radiological presentation where more frequent lesions should be ruled out first.

Keywords: solitary tumor; amyloidoma; spine; vertebrectomy; VCR

INTRODUCTION

Solitary amyloidoma (focal amyloid deposition) of the bone is rare. Most cases have been reported in the spine, mainly in the thoracic and cervical area [1-3]. It is a benign tumorlike lesion but characterized by local aggressivity with bone destruction and extension into the adjacent soft tissue, which in the case of the spine area, may result in vertebral instability and eventual spinal cord compression [4-6].

The exact etiology and pathogenesis of amyloidosis is unclear, however, amyloid deposits cause tissue destruction by progressive intercellular accumulation and pressure atrophy of adjacent cells.

CASE REPORT

A 52-years old man was admitted to the Emergency Department for middle back pain in association with neurological symptoms that had been evolving for three weeks.

Clinical examination showed sensitive impairment at the level of T5 and T6, ataxia, motor deficit in both lower limbs (4/5 motor strength), and pyramidal syndrome. Patient was classified as FRANKEL C. There were no anal or vesical symptoms. His general status was normal and there was no infectious context.

Radiologic assessment (MRI and CT-scan) showed solitary osteolytic lesion of T5 (Fig. 1 & 2) with spinal cord compression on the MRI due to epiduritis with invasion of the left pedicle and posterior arch. The lesion was in hyposignal on the T1 weighted sequence, with enhancement on contrast-enhanced sequence, heterogeneous and extensive like a malignant lesion (primitive or secondary).
Due to the neurological presentation, an urgent surgery was conducted. The procedure was the same as in other patients with vertebral metastasis consisting of a posterior approach with laminectomy of the concerned level (T5) and fixation from T4 to T6. Samples were taken from the surgical site for bacteriological and histological examination. The vertebral body of T5 was curetted as much as possible and cement was put to strengthen the body (Fig. 3).

**Figure 1 a & b.** MRI showing solitary osteolytic lesion of T5 with enhancement on contrast-enhanced T1-weighted images with posterior extension and spinal cord compression (epiduritis).

**Figure 2.** CT scan showing osteolytic lesion of T5 with posterior arch extension.

**Figure 3.** Postoperative control Xray showing the osteosynthesis from T4 to T6.
Results of histological examination confirmed the tumoral nature of the lesion and were compatible with a solitary amyloidoma of T5.

Clinical evolution was favorable with complete progressive recovery of the neurological symptoms. The postoperative course was uneventful with a patient without any pain at the 2 months follow-up visit.

Complete investigations were performed in order to look for a light chain myeloma, which is the most frequent disease responsible of amyloidoma. Salivary gland biopsy, blood exploration with protein electrophoresis, and biopsy of abdominal fat didn’t reveal myeloma.

After discussion with the oncologist, complementary treatment was decided and the patient underwent a course of radiotherapy one month after surgery.

At the six months postoperative visit, the patient came back to the outpatient clinic with middle back pain at the level of the surgical site. Clinical examination revealed no neurological symptoms at all. As the primary lesion was a solitary amyloidoma, full spine CT scan and MRI were performed to rule out a recurrence at the lesion site.

The new investigations showed no hardware issue but a local recurrence of the tumor at the level of T5 with local extension to T6.

Radiological aspect was similar to the one of the first admission with major development in the vertebral body of both vertebras and extension to the left pedicles and posterior arch on the left side (Fig. 4a, 4b and 5).

Given the pain presented by the patient and the wide recurrence of the solitary amyloidoma with high risk of new medullary compression, a new surgery was decided. An en-bloc resection of both vertebras T5 and T6 by an only posterior approach as described by Tomita [7] was performed.

**FIGURE 4 a & b.** CT scan six months after the first procedure. New lytic lesion at T5 with extension to the body of T6 including its left pedicle and posterior arch.

**FIGURE 5.** MRI of the thoracic spine (T2 Fat Sat weighted image) showing an extensive lesion at the level of T5 and T6 signing a recurrence of the solitary amyloidoma.
SURGICAL TECHNIQUE

The patient is positioned prone on four pads. After a midline incision is made, the paraspinal muscles are dissected subperiosteally from the spinous processes, lamina, and transverse process of the levels involved. Three spinal levels above and below the lesion are exposed to allow for instrumentation placement for fixation. The exposure is extended laterally to expose approximately 5 cm of the ribs lateral to the costotransverse joint at the level of the tumor and the levels immediately above and below the lesion, and the ribs are transected. Then, instrumented fixation by pedicle screws with the use of the free-hand technique is performed three levels above and below the diseased vertebrae to maintain spinal stability (T2 to T9 sparing T5 and T6). Meticulous blunt dissection is performed along the lateral wall to the anterior wall of the vertebral body, the great vessels are mobilized anteriorly away from the spine to avoid inadvertent vascular injury (Figure 6a).

Wide laminectomy is performed at the levels immediately above and below the lesion for wide exposure of the spinal canal. Opening of the foramen is performed bilaterally above and below the lesion. The pedicles are removed bilaterally, the nerve roots of the affected level are also ligated and cut on one side (left in our case) to provide enough space for mobilization of the vertebral body around the spinal cord.

Figure 6 a, b, c. Peroperative pictures
The thecal sac is dissected away from the venous plexus and the posterior longitudinal ligament. A temporary rod is placed on the right side. The anterior two thirds of the inferior endplate of the above vertebra and superior endplate of the below vertebra are cut using fine-threaded wires (Figure 6b). The cut is completed posteriorly by the osteotom with removal of the posterior longitudinal ligament. The vertebral body is detached from the spinal column and is removed en-bloc by mobilizing it gently around the spinal cord to the left side. Ventral reconstruction is then performed with a titanium mesh cylindrical cage, filled with autologous bone graft harvested from the iliac crest (Figure 6c). The instrumentation is completed by putting two definitive rods on both sides. Patient could stand at day 7 with a thoraco-lumbar orthosis for three months. Postoperative CT scan confirmed the total resection with good position of the cage (Figures 7a and b). Two years follow-up X-ray showed a stable construct (Figures 8a and b).

DISCUSSION

Few papers in the literature described solitary amyloidoma of the spine, with a controversy about its management, with articles defending a radical solution with total en-bloc resection because of the known risk of local recurrence [8], and others defending only intralesional resection with decompression and fixation with high chances of spontaneous resorption with time with good prognosis [9,10].


Detection of such a tumor is not easy due to its aspecific clinical and radiological presentation. Clinical presentation depends on the lesion location, size and involvement of neurological structures. Patient may be asymptomatic, having local back pain, or presenting partial or complete neurological deficits (paraparesis, paraplegia).

Definitive diagnosis is based on histopathology including immunohistochemistry for IgG lambda and
kappa light chains and the demonstration of amyloid protein A (AA) and anti-transthyretin obtained by open or percutaneous biopsy [9].

Twenty-six cases of solitary amyloidoma were reported in the spine literature. These articles described lesions involving the thoracic spine in fourteen cases, the cervical spine in nine cases (including three in C2) and three cases in the lumbar spine. Age at diagnosis varied between 38 and 79 years with average at 58.5 years with a majority of men. Half of the patients had neurological symptoms with spinal cord compression in 9 patients (70%), radiculopathy in 2 patients (15%) and cauda equina compression in 2 patients (15%) (Table I).

Hwang et al. [13] described the radiological aspect of this kind of lesion and recalled that MRI signal is the same as what can be seen in other locations of amyloidoma (lung, nasopharyngeal) with low-to-intermediate signal on T1-weighted images, intermediate-to-high signal on T2-weighted images, and variable enhancement on contrast-enhanced T1-weighted images.

Initial radiological findings should not primarily orient the clinician to such a diagnosis due to its rarity; it should be more likely focused on the diagnosis of metastasis or infectious disease. In the literature no author described specific MRI or CT characteristics to evoke this entity as a primary diagnosis.

Differential diagnosis includes locally aggressive processes like infections (Potts abscess), chondrosarcoma, lymphoma, plasmacytoma and metastases.

In regard to the management of amyloidoma, if no neurological signs are present with a stable spine, obser-

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<tr>
<td>BRIEF SUMMARY OF CASES REPORTED IN THE LITERATURE</td>
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<tr>
<td>Total</td>
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<tr>
<td>26 cases</td>
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<td>9 cervical (3 axis)</td>
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<td>3 lumbar</td>
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<td>13 Without neurological symptoms</td>
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<td>53.8% thoracic</td>
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<td>34.6% cervical</td>
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<td>11.5% lumbar</td>
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vation only is advised. If the patient shows neurological symptoms in relation to cord compression, some papers advised only decompression and stabilization, as progressive resorption over time will occur with excellent prognosis [10]. Other papers advocated a radical treatment of this tumor by a resection as complete as possible due to the risk of local recurrence [8, 14].

In the case of a vertebral resection, the main advantage of the posterior-only approach is that it allows for tumor excision and circumferential reconstruction of the spine in a single procedure; therefore, it reduces overall total operative time and perioperative morbidity rate.

The global prognosis depends on the presence or development of either a local recurrence or a systemic disease (breast, lung, gastrointestinal tract, central nervous system). An associated multiple myeloma should be excluded by bone marrow biopsy and immunoelectrophoresis of the serum and the urine as it changes the prognosis and requires a general treatment by chemotherapy [9].

In the case of our patient, intralesional resection was initially performed and patient regained neurological function of his lower extremities completely with normal bladder and bowel control. Surgical treatment was followed by radiotherapy. The role of radiotherapy in solitary amyloidoma of the bone has not been established, but seemed reasonable given the similar clinical and pathological features of amyloidoma and plasmacytoma. However, it did not prevent the occurrence of a recurrence, that compelled us to do a complete en-bloc resection of the affected levels during the second procedure.

CONCLUSION

Primary amyloidoma of the spine is very rare with sparse literature on the subject. It is considered as a benign lesion but it may engage functional prognosis or vital prognosis depending on its location and extension.

Combined intraluminal resection of the lesion and spinal stabilization is the treatment of choice, with no need for radical vertebrectomy initially, and recurrence rate is low. Careful follow-up of the patients is mandatory to ensure initiation of adequate systemic therapy if needed, or eventual secondary surgery with en-bloc resection.

REFERENCES