Solitary plasmacytoma of the spine: a 22 years follow-up case report

M. CAPPuccio, F. DE Iure, A. GASbarrini, S. BANDiera, S. Boriani

Unità Operativa di Ortopedia e Traumatologia – Chirurgia vertebrale, Ospedale Maggiore “C.A. Pizzardi” – Bologna (Italy)

Abstract. – Background and Objectives: Plasmacytoma is a systemic malignancy and it is the most frequent primary tumor affecting the skeleton. Progression from solitary plasmacytoma to systemic diffusion is reported to be 65-100% in 15 years. A case report of a rare 22-year follow-up of a thoracic solitary plasmacytoma is here presented.

Material and Methods: Clinical case analysis, radiographs, magnetic resonance images and histological sections of the lesion are discussed. Repeated surgical interventions were required due to progression and local recurrence of disease.

Results: Although the inadequate surgical treatment, low response to chemotherapy and radiation therapy, and many local recurrences, no systemic diffusion of the disease was observed in 22 years.

Discussion: Aggressive surgery may be indicated in young patients with isolated lesion and good prognostic factors. Palliative surgery remains necessary in order to decompress the cord and to stabilize the spine.

Key Words: Solitary plasmacytoma, Spine, Surgery.

Introduction

Myeloma is a primary and systemic malignant neoplasm which originates from the B lymphoid cells and is characterized by plasmacellular differentiation. It is the most frequent primary tumor affecting the skeleton. It is generally observed in males after 40-50 years of age. In 5-10% of cases an isolated bone localization, mostly in a vertebra, is present at onset and it’s commonly called “solitary plasmacytoma”. Although the course of the disease is rather unpredictable, a solitary plasmacytoma generally shows a systemic diffusion in 5-10 years from onset. What is remarkable in our case is that many local recurrences occurred without systemic diffusion of the disease all along a 22 year period. Progression from solitary plasmacytoma to multiple myeloma is reported to be 65-84% in 10 years and 65-100% in 15 years.

Case Report

In January 1984 a 36 years old male referred to a local hospital for an increasing back pain and progressive lower limbs weakness (Frankel C). Blood test were negative for infection or tumor while standard thoracic spine x-rays showed lytic changes in T9 and T10 (Figure 1). The patient underwent posterior laminectomy and tumor partial debulking improving his neurological status. In the following days the surgeons planned an anterior approach to complete the posterior debulking. Because of technical difficulties in isolating the thoracic aorta, the surgical procedure was not completed. Furthermore, despite the two operations, the pathologist only made a general diagnosis of malignant tumor. In the following weeks the patient was submitted to radiation therapy (36 Gy).

In October 1987, during a follow-up control, a progression of the tumor was observed and the patient underwent a new anterior approach by left thoracotomy for intralesional excision and fusion with autologous rib and metal rod instrumentation (Figure 2). At this time a diagnosis of plasmacytoma was achieved (Figure 3) and the patient was submitted again to radiation therapy (36 Gy). Although no signs of systemic diffusion of the disease were present, the patient was submitted to many cycles of chemotherapy (Melphalan 30-15 mg/day, Prednisone 50 mg/day) until 1992. The clinical and neurological status were excellent.

In January 1998, 10 years after the last operation, a new episode of increasing paraplegia oc-
curred and the patient lost the ability to walk. Imaging showed a new recurrence in T9 and T10 again without signs of systemic diffusion. The patient underwent immediate decompression and posterior T4-L2 instrumented fusion. Hystological diagnosis was well differentiated plasmacytoma. No adjuvant therapies were done and the neurological status recovered again.

Six years later, in October 2004, the patient began to complain inferior limbs weakness and numbness. MRI showed a new recurrence (Figure 5) while immuno-electrophoresis and urinary electrophoresis remained negative. In January 2005, because of a quick neurological worsening, the patient had a new posterior surgical revision with a complete decompression of the spinal cord. Functional recovery was slower this time, nevertheless in 6 months the patient was able to walk with supports.

The last clinical and radiological control in January 2006 showed no local recurrence and nearly complete neurological recovery (Figure 6).

**Discussion**

Multiple myeloma is a malignant tumor of the bone marrow characterized by plasmacellular differentiation mainly affecting the skeleton. Favorite sites of onset in the skeleton are those rich in bone marrow and, particularly, vertebral bodies more often than pedicles and posterior arches. Skeletal changes go through different steps such us trabecular infiltration, osteolysis due to osteoclasts activation, tumor mass formation. A solitary lesion is found in 5-10% of multiple myeloma at onset. Histopathologic features in solitary...
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Figure 2. A, B. Thoracic spine plain x-ray after second anterior excision and fusion.

Figure 3. Gross pathology following the second anterior excision in 1987.

Figure 4. Thoracic spine MRI showing local recurrence in T9-T10 in 2005.
plasmacytoma and multiple myeloma are the same showing the same disease affecting B lymphoid cells. Nevertheless from a clinical point of view, clinical course and prognosis in solitary plasmacytoma and multiple myeloma are different. Solitary plasmacytoma of the bone, particularly in spine locations, has a better prognosis and tends to affect elderly people. Besides these differences most authors consider radiation therapy the best treatment of spine locations in both types. Chemotherapy is generally associated depending on the systemic diffusion of the disease. Clinical course of plasmacytoma, following radiation therapy, depends on initial stag-

Figure 5. Gross pathology following the last surgical procedure is the same as in 1987.

Figure 6. A, B. Thoracic spine standard x-rays taken at the last control in 2006.
ing and other prognostic factors such as tumor mass or persistent of monoclonal gammopathy. However, half of solitary plasmacytomas turn into multiple myeloma 2 to 10 years from diagnosis (average 3.5 years)16-18. Modern therapy of multiple myeloma relies on chemotherapy. Thanks to the last 20 years improvements in therapeutic strategy, the median survival of these patients increased of 2-3 years. Particularly contributed to these results high dose chemotherapy (Melphalan 140 mg/m²)19,20 followed by bone marrow transplantation and new drugs specifically active in the marrow micro-environment20.

The role of surgery in the treatment of spine lesions from plasmacytoma or multiple myeloma is not completely defined yet. Commonly accepted surgical procedures are those aiming to decompress cord, reduce pain and stabilize spine in order to allow weight bearing without external orthosis. From an oncological point of view surgery can be employed to reduce or remove a tumor mass if it is low sensitive to radiation therapy and chemotherapy11. Therefore the main role of surgery in systemic diseases affecting the spine is to improve the quality of life of patients also by a local control of the tumor mass.

In the treatment of spine metastasis, in very selected cases (solitary lesion, primary treated, good prognosis), the best local control can be achieved by an “en bloc” excision with oncologically adequate surgical margins; in our case review we had no local recurrences in patients submitted to this treatment21.

In the reported case, where we counted many local recurrences during more than 20 years without systemic diffusion, an “en-bloc” excision could have avoided many repeated palliative surgeries, although they were effective from a clinical point of view. In the literature we found evidences that a wide margin en bloc resection is successfully adopted in young patients with solitary plasmacytoma and good prognosis factors7,22,23. Nevertheless, the indication to such an aggressive surgery in a disease sensitive to radiation therapy and chemotherapy is not well defined yet, and vertebrectomy is commonly considered an overtreatment. The critical point would be in recognizing a solitary lesion with no tendency to systemic diffusion. In such case a surgical excision could also reduce or avoid the need for adjuvant therapies rich in side effects.

In conclusion, aggressive surgery could be indicated in young patients with isolated lesion and good prognostic factors. Palliative surgery remains necessary in order to decompress the cord and to stabilize the spine. Radiation therapy alone should be employed in those cases in which surgery is technically not feasible.

References

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