Case Report

Transverse syndrome as the dominant feature of solitary cervical plasmocytoma with dissemination

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Study design: A case report of transverse syndrome secondary to compression from an intravertebral plasmocytoma.

Objectives: To report a rare case of cervical cord compression.

Setting: Vienna, Austria.

Case report: A 47-year-old man presented with pain over the left scapula and subsequent development of pain and sensory disturbance. Investigations showed that the entire C7 vertebra was affected by a plasmocytoma. Decompression was carried out but no stabilisation of the spinal column was performed. Seven days after surgery the patient became paraplegic. Chemotherapy produced no improvement.

Conclusion: Transverse syndrome may be a dominant feature of plasmocytoma and may resolve after immediate decompression but may recur if the spinal column is not stabilised.

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Keywords: plasmocytoma; malignancy; neurosurgery; back pain; chemotherapy

Introduction

Transverse syndrome due to compression of the cervical spinal cord by an expanding, intravertebral plasmocytoma is rare.^{1–4} Usually, spinal plasmocytomas are located in the thoracic and lumbar vertebra.^{5–10} We present the data of a patient with intravertebral plasmocytoma at C7 with bone marrow dissemination, who presented with back pain for 4 months and transverse syndrome for 1 month prior to diagnosis.

Case report

The patient is a 47-year-old, well trained rock climber and mountain biker who started to complain about dull pain over the left scapula since June 2000. The pain was relieved by warmth and deteriorated in cold weather. Despite therapy with ultrasound, the pain started to radiate towards the left shoulder and left upper arm. Diclofenac was effective so the patient was able to partially continue his sporting activities. From early September 2000, numbness of the hands and lower arms occurred. Simultaneously, the muscles became sore. A few days later, he noticed increasing

weakness of both lower limbs and increasing tension of the abdominal muscles. In the middle of September 2000 his gait became evidently disturbed and he tired easily. Blood sedimentation rate, blood cell counts, blood glucose, urea nitrogen, creatine, uric acid, serum lipids, γ -glutamyl-transpeptidase, alkaline-phosphatase, rheuma factor, C-reactive protein, anti-streptolysin-O-titer were normal at that time. X-ray of the lungs revealed an elongated aortic arch but no other abnormality. ECG showed sinus bradycardia (49 beats/minute). Transthoracic echocardiography and ultrasonography of the abdomen were not informative. Nerve conduction and F-wave studies of the left and right peroneal and tibial nerves, were normal, as well as conventional needle EMG of the right lateral vastus muscle. Plain radiography of the spine and pelvis showed hyperostosis of vertebra C6 and C7, reduction of the intervertebral space C6/7, spondylosis deformans, increased thoracic kyphosis and wedge-shaped destruction of some thoracic vertebrae. In October 2000 the patient was first seen by a neurologist.

Clinical neurological examination on admission revealed distal weakness (Medical Research Council (MRC) grade 4+), and wasting of both upper extremities, a reduced biceps tendon reflex on the left

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Figure 1 Sagittal T2-weighted MRI (TR/TE 5000/132) of the cervical spine reveals replacement of the bone marrow by a hypointense, intravertebral tumour C7 that protrudes circumferentially around the vertebral canal, involving the pedicles, the arches and the processus spinosus (A). Sagittal T1-weighted KM enhanced (Magnevist® 30 ml) (TR/TE 600/15) images additionally show that the tumour penetrates the vertebral cortex, has a soft tissue component that compresses the spinal cord, and affects the adjacent intervertebral discs and vertebra (B). An axial T1-weighted KM-enhanced image (TR/TE 500/15) shows the markedly reduced CSF-space (C)

side, diffuse weakness of both lower limbs (MRC grade 4), subclonic patella tendon reflexes, a positive Babinsky's sign bilaterally, and a sensory level at Th3. Romberg's test revealed slight imbalance. On blood chemical investigations creatine was 1.4 mg/dl (normal: 0.5–1.2 mg/dl), and total protein 8.2 g/dl (normal: 6–8 g/dl). Serum albumin was 48.3% (normal: 55–70%), β -globulin 7.7% (normal: 8–14%), and γ -

globulin 34% (normal: 10–20%). Serum IgG was 2700 mg/dl (normal: 700–1600 mg/dl) and κ -light chains 906 mg/dl (normal: 170–370 mg/dl). There was no abnormal protein on immunofixation. CSF albumin was 79.7 mg/dl (normal: <35 mg/dl), IgG 25.3 mg/dl (normal: <3.4 mg/dl), and IgA 1.4 mg/dl (normal: <0.5 mg/dl). Magnetic resonance imaging (MRI) of the cerebrum was normal. MRI of the

cervical spine showed high grade vertebral stenosis at C7 due to a space-occupying process within the axially and sagittally enlarged vertebrum C7, including the dorsal vertebral arch, the processus spinosus, and neighbouring segments (Figure 1). The pre- and retro-medullary space at C7 was completely reduced with partial, mainly posterior compression of the spinal cord. Immediately after the spinal MRI, the patient developed anaesthesia below Th3, urinary retention, and lost his ability to walk due to severe lower limb paraparesis (MRC grade 0 on the left and grade 1 on the right side). Immediately he underwent surgical decompression by complete laminectomy C7 (posterior decompression), and partial resection (intralesional excision) of the tumour (anterior decompression). No auto- or allograft material was implanted to fill the defect, and no stabilisation of the spinal column was performed. Histological examination of the tumour revealed plasmacytoma grade 2 (Figure 2). One day after surgery, the patient was able to move his legs again, and 1 day later he could walk with support. Pain had resolved to such a degree that analgesics were unnecessary. A CT scan of the skull showed multiple osteolytic lesions and iliac bone marrow aspiration revealed 6% malignant plasma cells. Seven days after surgery, he suddenly became paraplegic. Neuroimaging revealed acute cord compression at C7. This time the cervical spine was extended by means of a halo cast. Additionally, chemotherapy with vincristine, doxorubicin and dexamethason (VAD regimen) was begun without improvement.

Discussion

Plasmacytoma is a tumour composed of plasma cells that organise in one or more solitary tumour(s) in the

Figure 2 Histological examination of the abnormal tissue after surgical removal from vertebra C7 shows sheets of tightly packed atypical plasma cells. The cells have round or oval, often two or more eccentric nuclei, clumped chromatin, sometimes visible nucleoli, and abundant basophilic cytoplasm with a prominent paranuclear pale area (Haematoxylin & Eosin, $\times 400$)

skeleton (osseous plasmacytoma), in the soft tissue (extramedullary plasmacytoma), or seed diffusely throughout the bone marrow (multiple myeloma).¹¹ The distinction between solitary plasmacytoma and multiple myeloma is important as solitary plasmacytoma is very radiosensitive with the theoretical chance of cure, following early diagnosis.⁶ Usually, plasmacytomas produce a monoclonal immunoglobulin in blood, urine, or both (M-component), and cause lytic bone lesions.¹² In 3–7% of the cases, plasmacytoma occurs as a single solitary bone tumour.^{3,5,6,8,9,13} Among these, 25–60% occur within the spine, most frequently in the thoracic and lumbar vertebrae.^{5,8} Solitary vertebral plasmacytoma may include the vertebral body alone or additionally the posterior elements. Solitary plasmacytoma may lead to collapse and compression of the vertebral body and endplate fractures.³ In rare cases the tumour penetrates the vertebral cortex and develops a soft tissue component that may involve the intervertebral disc and adjacent vertebra and may lead to cord compression.^{3,11} Cord compression occurs in two-thirds of the patients with solitary vertebral plasmacytoma, but only in 7–16% of the patients with multiple myeloma.⁸ Patients with solitary vertebral plasmacytoma present with back pain, radicular pain, sensory disturbances, and para- or tetraparesis.^{5,8} There is a preponderance in men.^{5,6} The mean interval between onset of symptoms and diagnosis is 1 year.^{5,8} Staging of plasmacytoma is necessary for optimising therapeutic strategies, and relies on the number of lytic bone lesions, the amount of serum haemoglobin, serum calcium, and monoclonal immunoglobulins.¹² These variables are used because they have been shown to correlate with the tumour mass and survival time.³ A good prognostic factor of solitary plasmacytomas is the absence of an M-component, found in half of the patients.⁹ Patients with an M-component have a higher risk of dissemination and local recurrence of the tumour than patients without.^{5,8} Despite the more favourable prognosis of single compared to multilocal solitary plasmacytoma, both may evolve into multiple myeloma within 3–5 years.^{5,9,13} Multiple myeloma appears to be inevitable if a solitary osseous plasmacytoma is given enough time without treatment.⁸

In the presented patient the diagnosis of plasmacytoma was based on the presence of an M-component, increased IgG in the serum and CSF, increased serum kappa light chains, histological examination of the spinal tumour, and the bone marrow aspiration. Initially, the tumour was regarded as a solitary osseous plasmacytoma, but bone marrow examinations and radiography of the skull after surgery demonstrated systemic bone marrow infiltration and multiple osteolytic lesions in the skull. The presented patient is noteworthy because he is young, the initial symptom was pain over the left scapula, the dominant clinical feature was transverse syndrome, the diagnosis was established 4 months after onset, the entire C7 vertebra, including body and posterior elements with

the processus spinosus, was affected, and the adjacent discs and vertebra were involved. In this patient it is quite likely that initially there was a solitary, cervical plasmacytoma, which became symptomatic after it had disseminated and turned into multiple myeloma.

Therapy of plasmacytoma comprises radiotherapy, surgery, and chemotherapy.⁵ The primary treatment of solitary plasmacytoma is local irradiation.^{5,11} Local irradiation is highly effective in achieving local control over the tumour and palliation.⁶ The usually recommended dose is 40–50 Gy, given in daily fractions for 3–4 weeks.⁸ Higher dosages are not feasible because of spinal cord toxicity.⁸ Surgery is useful for decompression in case of cord compression or other neighbouring structures, and for histological diagnosis.^{5,11} Surgery comprises posterior (laminectomy) or anterior decompression (anterior procedure) with total or partial resectioning or curettage of the tumour.⁸ Decompression should be associated with reconstruction, fixation and stabilisation by osteosynthesis.¹ In the presented patient bone reconstruction and fixation were not performed, because there was only partial resectioning and the local situation was regarded sufficiently stable. Adjuvant chemotherapy has no established place in the management of solitary plasmacytoma since it does not prevent the development of multiple myeloma.^{8,9} However, prophylactic therapy with interferon- α may delay the evolution of multiple myelomas in patients at high risk of dissemination.¹³ A positive effect of interferon- α combined with alkylating agents remains so far unproved.¹³ Contrary to solitary plasmacytoma, multiple myeloma is treated only with chemotherapy. For chemotherapy, melphalan-prednisone, the VAD regimen, high-dose dexamethasone, high-dose alkylating agents, and myeloblastic therapy plus autologous cell transplantation are proposed.⁸

In conclusion, this case shows that transverse syndrome may be the dominant feature of intravertebral, cervical plasmacytoma with diffuse skeletal dissemination. Transverse syndrome may resolve after immediate decompression by laminectomy and tumour

resectioning, but may recur if the spinal column is not stabilised.

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