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Spinal Leptomeningeal Infiltration by Systemic Cancer: Myelographic Features

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Five cases of spinal leptomeningeal infiltration by systemic cancer are presented and myelographic features are discussed with a review of the literature. Common characteristic features are parallel longitudinal striations due to thickened nerve roots in the cauda equina and bizarre irregular filling defects with varying degrees of blocks, resembling arachnoiditis. Another pattern described in the literature consists of multiple nodular filling defects along the nerve roots of the cauda equina.

Nonneurogenic leptomeningeal infiltration by systemic cancer is a rare mode of metastasis to the central nervous system. This process is characterized by diffuse, sheetlike infiltration of the pia-arachnoid. Symptoms and signs are produced by meningeal irritation, increased intracranial pressure or neuronal dysfunction, either cerebral or spinal, or a combination of these elements [1]. The syndrome has been known since the turn of the century. The spinal form of the disease is not as well known and less common than the cerebral form [2]. Although this entity is still considered rare, its frequency is increasing, especially with systemic lymphoma [3-5]. Clinical manifestations of spinal leptomeningeal infiltration are obscure and nonspecific and an early diagnosis is often difficult [2, 3]. The myelographic description of this type of spinal cord and cauda equina malignancy has been infrequently reported. We present five cases with a review of the literature.

Case Reports

Case 1

A 48-year-old man was found to have undifferentiated lymphoma involving the small bowel, left ureter, and left axillary and submandibular nodes 5 months before admission. He received radiation therapy to the abdomen and was treated with chemotherapy. He was admitted after 3 weeks of progressive pain in the low back and both thighs. He also noted weakness in going up and down stairs over the previous several days. Physical examination showed weakness of the iliopsoas muscles and diminished muscle stretch reflex in the lower extremities. There was no sensory abnormality.

At the time of myelography, the spinal fluid was viscous and xanthochromic. The protein content of the spinal fluid was 4,400 mg/dl and the cytology was negative. The myelogram showed thickened nerve roots with parallel longitudinal striations involving the entire lumbar region (figs. 1A and 1B). Radiation therapy to the whole spinal axis was immediately initiated. His symptoms and signs continued to worsen rapidly.

A laminectomy was performed at the T12-L2 level. The dura appeared normal and the underlying arachnoid was also grossly normal. However, there were a few small whitish areas in the arachnoid. The roots of the cauda equina appeared swollen and somewhat hyperemic. The biopsy specimen of the arachnoid showed diffuse infiltration with lymphoma cells in the arachnoid (fig. 1C). Despite radiation and chemotherapy, the patient continued to deteriorate with third nerve palsy and right arm paralysis. He died 2 weeks later.
Case 2

A 61-year-old man was hospitalized with complaints of low back pain radiating to both legs and difficulty in walking of 2 weeks duration. He was found to have hilar, submandibular, and gastric masses 2 months before admission. The biopsy from the gastric mass showed histiocytic lymphoma. The physical examination revealed a partial left seventh nerve palsy, decreased muscle strength in the hip extensors bilaterally, and reduction of pinprick sensation in both heels. A myelogram was obtained (figs. 2A and 2B). The spinal fluid was viscous and xanthochromic. The protein content of the spinal fluid was 2,400 mg/dl. Lymphoma cells were identified in the spinal fluid (fig. 2C). The myelogram showed diffuse thickened nerve roots with parallel longitudinal striations. Radiation therapy to the whole spinal axis was immediately initiated.

Case 3

A 68-year-old woman was admitted because of confusion and memory loss. She had a right radical mastectomy for breast carcinoma 15 years before and left breast carcinoma was discovered 2 months earlier. Computed tomography (CT) of the brain showed extraventricular obstructive hydrocephalus. A ventriculoperitoneal shunt tube was inserted. Cerebrospinal fluid analysis showed class IV adenocarcinoma cells. She received radiation therapy to the brain and intrathecal methotrexate through the reservoir of the shunt. Her condition improved and she was discharged. She was readmitted 4 months later because of difficulty in walking.

Physical examination revealed weakness of the lower extremities, reduction of position sense of the toes, and absence of deep tendon reflexes in the lower extremities. Repeat CT of the brain showed no evidence of metastasis. Ventricular size was within normal limits. A total myelogram was obtained. The spinal fluid was xanthochromic. The protein content of the spinal fluid was 880 mg/dl and the cytology showed class IV adenocarcinoma cells. The myelogram showed markedly thickened nerve roots with parallel longitudinal striations in the cauda equina (figs. 3A and 3B). Radiation therapy to the entire spinal axis was immediately initiated.
Case 4

A 45-year-old man developed left eye proptosis without loss of visual acuity. Tissue surgically removed from the left retrobulbar space 1 year later was diagnosed as pseudotumor. He underwent radiation therapy with good response. Left forehead and left neck masses developed 2 years later. Excisional biopsy of the right supraclavicular lymph node revealed numerous atypical plasma cells. Serum protein electrophoresis demonstrated a monoclonal IgM spike, but the bone marrow biopsy was negative. Radiation treatment of presumed extramedullary plasmacytoma obtained a good response.

The patient was hospitalized 7 years later after 6 months progressive weakness in the lower extremities. He complained of some pain in both quadricep and hamstring areas. On physical examination, he had decreased strength in the proximal muscles and distal hypoesthesia. Myelography showed thickened nerve roots in the cauda equina with parallel longitudinal striations (fig. 4). Cerebrospinal fluid protein was 325 mg/dl; glucose, 60 mg/dl; and monocytes, 39%. Cytology was negative. The patient underwent radiation treatment of a total tumor dose of 1,800 rad (18 Gy) to the whole craniospinal axis. At the end of treatment, he was pain free with some improvement in strength in the lower extremities.

Discussion

Leptomeningeal infiltration by systemic cancer is characterized by diffuse or multifocal infiltration of the pia-arachnoid prolongations around cranial and spinal nerve roots and into the perivascular spaces of the superficial cortex. This form of tumor growth is distinct from the metastatic nodular proliferations of the parenchyma and dura. The tumor grows in sheetlike fashion along the surface of the brain and spinal cord, at times eliciting an inflammatory reaction. It is this pathologic picture that is termed meningitis carcinomatosa. Gross inspection of the brain, spinal cord, and nerve roots often reveals little or no abnormality. Among the positive gross findings, leptomeningeal thickening is commonly described as opaque, opalescent, milky, and whitish. Discrete nodules or focal granularity may be seen,
attributed to focal accumulation of proliferation of cancer cells in the leptomeninges. These nodules are most often found along nerve roots of the cauda equina [1–3].

The mode of tumor spread to the leptomeninges has been controversial. Peripheral lymphatic invasion, hematogenous dissemination, seeding through the choroid plexus, or spreading of the tumor from the brain parenchyma and nerve roots to the cerebrospinal fluid have been suggested [1, 3, 4].

Van Allen and Rahme [6] reported a case of diffuse leptomeningeal lymphosarcomatous infiltration of the cauda equina with myelographic appearance of longitudinal parallel striations. At surgery, markedly swollen nerve roots of the cauda equina appeared like a mass of "spaghetti." The myelographic features of our cases 1–3 and 5 were identical. A similar pattern was observed by others in cases of lung carcinoma and melanoma [7–9]. The parallel striations correlated with thickened nerve roots by diffuse tumor infiltration. The cauda equina and lumbosacral region are the most common sites of spinal involvement. The infiltration is more on the dorsal surface, probably due to gravitational effect [1, 2].

Guyer and Westbury [8] noted multiple nodular filling defects along the nerve roots of the cauda equina in a case of lung carcinoma. Similar findings were described by others [3, 10, 11]. The multiple nodules correlate with focal accumulation or local proliferations of cancer cells in the leptomeninges along the nerve roots.

Prentice et al. [10] described a complete block of Pantopaque in the lower thoracic region in a case with reticulum cell sarcoma. The contrast material was introduced via cisternal puncture after unsuccessful lumbar punctures. At the level of the block, the contrast column was irregular, resembling that of chronic adhesive arachnoiditis. Case 5 demonstrated a similar complete block with irregular margins at the thoracolumbar junction in addition to an almost complete block at the L4–5 level (figs. 5A and 5B).

Lumbar puncture is often difficult to perform because of obliteration of the subarachnoid space due to swollen nerve roots, occasionally resulting in dry puncture. It may cause severe radiating pain to the lower extremities. The spinal fluid is often xanthochromic and occasionally viscous due to increased protein content. The cytology is positive in 20%–50% of cases [1–3].

Oil contrast material (Pantopaque) was used in all of our cases. Ree et al. [11] advocated Amipaque, demonstrating multiple nodular defects along the nerve roots in the cauda equina in a case of spinal leptomeningeal carcinomatosis. The swollen nerve roots would probably be demonstrated to better advantage by using a water-soluble contrast material. However, since the lesions are poorly localized clinically, a total myelogram is often requested, in which case we prefer using Pantopaque. Even in patients with suspected spinal metastasis whose clinical findings suggest a lesion in the lumbar region, it is important to evaluate the entire spinal canal in search of other clinically silent lesions. In cases with blocks in the cervical or upper thoracic region, the tumor margins are usually better demonstrated with Pantopaque. Another advantage of using Pantopaque is that some of the contrast material may be left in the spinal canal for future follow-up, if necessary.

In summary, one of a combination of the following myelographic patterns has been observed in the spinal leptomeningeal infiltration by systemic cancer: (1) parallel longitudinal striations in the cauda equina due to thickened nerve roots; (2) bizarre, irregular filling defects with varying degrees of blocks, resembling arachnoiditis; or (3) multiple nodular defects along the nerve roots of the cauda equina.

The main differential diagnostic possibilities are hypertrophic interstitial polyneuritis [12] and arachnoiditis. Differentiation of the parallel longitudinal striations in the cauda equina from hypertrophic interstitial polyneuritis may be impossible. If there is associated irregularity of the contrast column in addition to the longitudinal striations, hypertrophic
interstitial polyneuritis is less likely since the arachnoid lining should be smooth. However, the combination of longitudinal striations and irregularity of the contrast column may be difficult to differentiate from arachnoiditis [13].

Differentiation of irregular filling defects with varying degrees of block from arachnoiditis may also be impossible. However, in a clinical setting when the spinal form of leptomeningeal infiltration by systemic cancer is suspected, one or a combination of the above myelographic patterns will be helpful in establishing the diagnosis.

REFERENCES