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Case Report

Pseudomyxoma Peritonei Lesion of the Thoracic Spine: A Case Report and Literature Review

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Abstract

Background: Pseudomyxoma peritonei (PMP) is a rare condition, with no previously described location in the spine. Here is presented a case of PMP extension to the thorax and then the spine, treated with two-level vertebrectomy, laminectomy, and posterior fusion.
Case Report: We report here the case of a 64-year-old man presenting pathological fractures of T6 and T7 due to PMP extension to the pleural cavity and thoracic spine. He presented interscapular pain with mild spinal cord compression symptoms. He was treated by achieving T5-T8 laminectomy, T2-T10 pedicle fixation, and T6-T7 vertebrectomy. The postoperative course was uneventful apart from a pulmonary embolism (PE) with favorable evolution under anticoagulants.
Conclusion: To achieve spinal cord decompression, kyphosis correction, and spine stabilization, a two-level vertebrectomy, laminectomy, and posterior fusion would be beneficial for a PMP lesion of the thoracic spine.

Keywords: Pseudomyxoma Peritonei; Disease; Spine

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Background

Pseudomyxoma peritonei (PMP) – also called "gelatinous disease of the peritoneum" or "gelatinous ascites of the peritoneum" –is a rare disease. It was first described in 1842 by Karel Rokitansky and was officially named "pseudomyxoma peritonei" in 1884 by Richard de Werth (1, 2). Perforation and dissemination phenomena of a mucin-producing tumor, mainly from appendiceal mucocele or ovarian adenocarcinoma origin, lead to a diffuse intraperitoneal accumulation of gelatinous mucinous lesions (3). Frequently symptomless in the early stages, beginning with vague abdominal symptoms, it is not rare that PMP is diagnosed as an unexpected finding during an abdominal procedure for suspected appendicitis or ovarian mass (4, 5). Appendicitis with increased abdominal girth and a fluid-filled appendix with curvilinear calcifications on computed tomography scan (CT-scan) images may orientate the diagnosis towards PMP (6).

Most PMP cases are classified as low-grade, about 78%, and rarely spread through the bloodstream or lymphatic system (7). Nevertheless, although PMP is usually considered a slowly growing benign lesion, rapidly progressing invasive diseases are described. Consequently, PMP should be considered as a "borderline malignant" process. Four major treatment strategies are available: symptomatic management, chemotherapy, surgery, and rarely radiotherapy due to the unbearable side effects of such large-field tumors (4).

Extra-peritoneal spreading of PMP is rare, and spinal localization has never been described in the literature so far (8, 9). This study aimed to report a patient presented with a T6-T7 pathological fracture secondary to a PMP lesion, treated by total vertebrectomy.

Case Report

A 64-year-old man was diagnosed in 2009 with PMP originating from the appendix. He was treated with debulking surgery and intraperitoneal chemotherapy. Disease evolution was marked by a recurrence in 2021 at the pleural and pericardial levels, requiring surgical procedures. A left pneumonectomy was performed, with a recurrent laryngeal nerve palsy sequela.

He was referred to the orthopedics unit, presenting with interscapular pain associated with semi-belt neuropathic pain at the T6 level on both sides. Upon clinical examination, a significant regional kyphosis in the mid-thoracic region was noted without any neurological deficit. However, his reflexes were sharp on the left lower limb and normal on the right side. No other signs of pyramidal irritation, sphincter disorder, or proprioception anomaly were found.

Imaging Features

Preoperative CT-scan showed a pathological fracture of T6 and mainly T7 with an important kyphosis (35° regional kyphosis) and vertebral body collapse bulging into the canal (Figure 1).

Axial images showed extensive gelatinous masses presenting low attenuation involving the left pleura, lung, and adjacent vertebral body. Curvilinear and peripheral calcifications, as frequently seen in this condition, were also observed around the vertebral bodies of T6 and T7 in the axial plane (Figure 1).

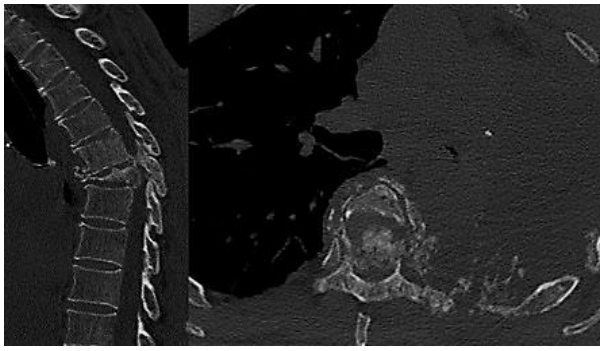

Figure 1. Preoperative computed tomography (CT) scan in sagittal view (on the left) showing kyphotic fracture of T6 and mainly T7. The axial view (on the right) shows extensive low-attenuation gelatinous masses involving the left pleura, lung, and adjacent vertebral body.

Axial images showed extensive gelatinous masses presenting low attenuation involving the left pleura, lung, and adjacent vertebral body. Curvilinear and peripheral calcifications, as frequently seen in this condition, were also observed around the vertebral bodies of T6 and T7 in the axial plane (Figure 1).

The preoperative magnetic resonance imaging (MRI) exhibited a 25cm-long left latero-thoracic mass invading T6 and T7 vertebral bodies and transverse processes, the posterior adjacent ribs, and the medullary canal (Figure 2a). This mass occupied almost the whole pneumonectomy area on the left side from the chondro-sternal junction to posterior soft tissues, with intimate contact with the pericardium and the descending thoracic aorta and extended to the retroperitoneum. T2-weighted axial MRI image demonstrated extensive multiseptated mass involving both left pleura, lung, and adjacent vertebra. This mass appeared in high signal on T2-weighted sequences due to their mucin composition. Contrast-enhanced T1-weighted axial image demonstrated areas of slight enhancement within the lesions as usually seen in mucinous tumors (Figure 2b).

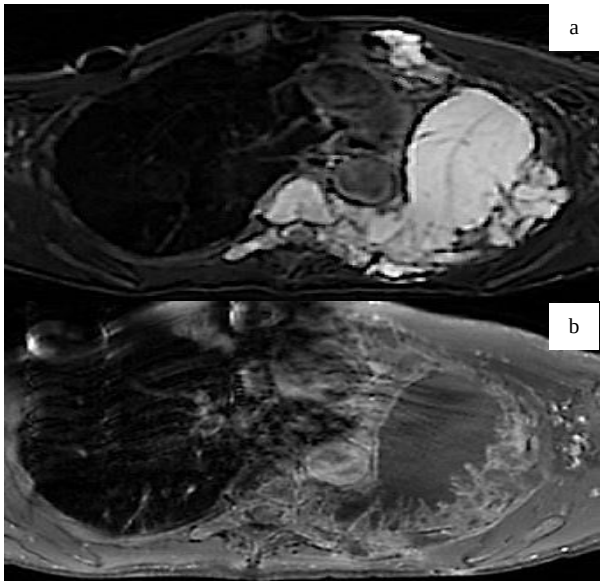

Figure 2. a: T2-weighted axial magnetic resonance imaging (MRI) demonstrated extensive multiseptated mass involving both left pleura, lung, and adjacent vertebra. They appear in high signal on T2-weighted sequences due to their mucin composition; b: Contrast-enhanced T1-weighted axial magnetic resonance imaging (MRI) exhibiting areas of slight enhancement within the lesions as usually seen in mucinous tumors

Surgical Intervention

After being clearly and well informed of the risks and benefits of the surgery, the patient accepted the proposed surgical procedure of T6 and T7 vertebrectomy. The goals of this procedure were to decompress the spinal cord, correct regional kyphosis, and stabilize the spine. Under general anesthesia, in the prone position with a Mayfield skull clamp, a median posterior approach was performed to expose the thoracic spine from T2 to T10. Pedicle screws were placed bilaterally in the overlying and underlying vertebrae above, from T3 to T5 and T8 to T10. Spinal exposure was then extended laterally to expose the costal arches from T5, T6, T7, and T8 on the left side, exhibiting characteristic gelatinous lobules (Figure 3). The tumoral invasion of the intercostal spaces led directly to the pneumectomy cavity. A large amount of gelatinous tumor was removed. A complete laminectomy from T5 to T8 was then performed.

Figure 3. Intraoperative picture showing the gelatinous lobules on the left side of the thoracic spine (white dotted circle)

T6 and T7 nerve roots were then sectioned bilaterally to approach vertebral bodies up to their anterior surfaces. Starting from the left side with a temporary rod on the other side, T5-T6, T6-T7, and T7-T8 discs were resected. Vertebral bodies were removed using an ultrasonic bone scalpel. A complete T6 and T7 vertebrectomy was then performed by completing the resection from the other side (10). The spinal cord was thus gradually decompressed. There was also tumor invasion along the aorta in the front. The maximum tumor volume was resected; however, it was not a complete resection. A corpectomy mesh cage, filled with an autologous bone graft from laminectomy, was then placed between the vertebral bodies of T5 and T8. The two final rods were then placed by correcting local kyphosis, and additional bone grafting was performed on the transverse processes. Abundant washing with physiological saline and closure plan by plan on suction drainage was done. The total operating time was 7 hours.

Pathology Findings

Expectedly, gelatinous mucinous lesions were found. All samples were occupied by tumoral extension, with patches of acellular mucus on most samples. Epithelial flaps were associated with these lesions, around or within mucus patches. The vertebral bone tissue was broadly replaced, mainly by abundant acellular mucin with the scanty lining of mucinous epithelium showing no or little cytologic atypia (Figure 4a, b, and c).

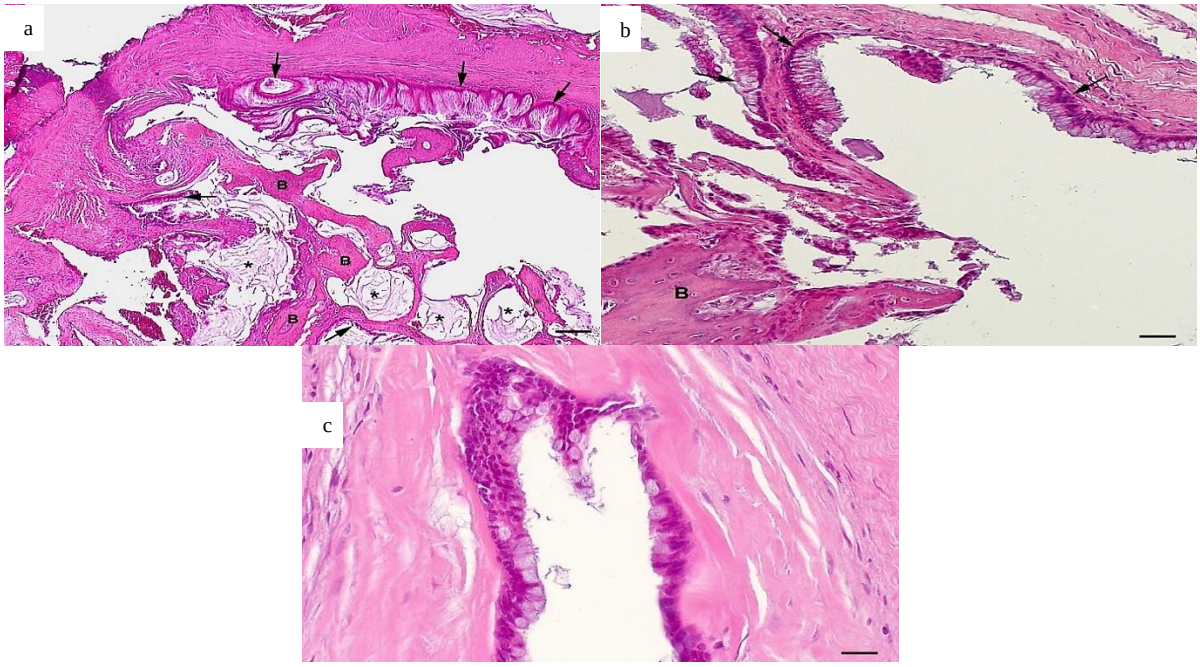

Figure 4. a: The bone tissue (B) is broadly replaced by pools of acellular mucin (asterisks). A mucinous epithelium (arrows) is lining some mucinous cavities. [hematoxylin and eosin (H&E) stain, scale bar = 75 µm]; b: The mucinous epithelial lining (arrows) is most often regular without cellular atypia. [B: Bone, hematoxylin and eosin (H&E) stain, scale bar = 20 µm]; c: In some spots, the mucinous epithelium lining shows low-grade cellular atypia. [hematoxylin and eosin (H&E) stain, scale bar = 10 µm]

It corresponds to a pattern of a low-grade mucinous neoplasm metastatic from a primary appendix tumor (11).

Postoperative Course

After surgery, back pain was moderate under analgesic treatment with no radicular pain nor neurological deficit in the lower limbs. Postoperative X-ray confirmed the adequate position of the implants and kyphosis reduction (Figure 5). A pulmonary embolism (PE) was diagnosed on day five and treated with three months of anticoagulation. The CT-scans performed at six months and one year were satisfactory, showing no lesion recurrence at the operated level.

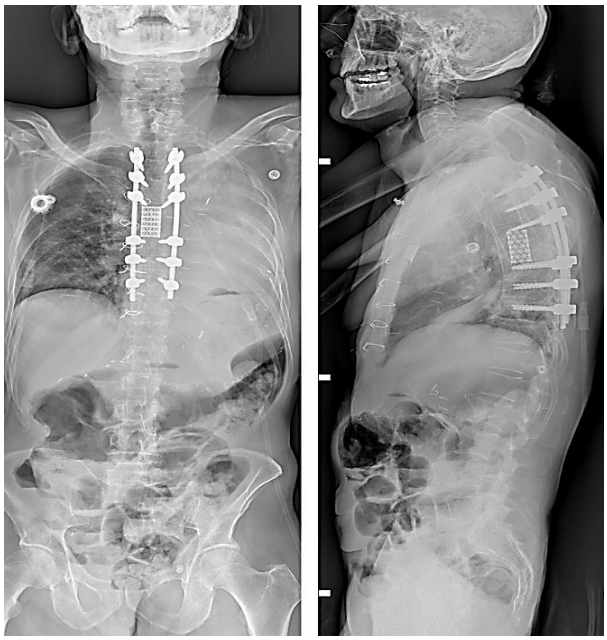

Figure 5. Postoperative standing full-spine X-ray showing the adequate position of the implants and kyphosis reduction

Discussion

Smeenk et al. reported a PMP incidence of approximately 2 cases per million inhabitants per year, with the appendix as the main origin site (82%) (12). However, the primary location site can also be the ovary (13).

PMP was also reported in patients diagnosed with rectal cancer and in patients with an extraperitoneal iliopsoas abscess secondary to mucinous adenocarcinoma of the appendix (14, 15). Extra-abdominal locations of PMP are exceedingly rare and can occur synchronously or several months after initial diagnosis (16). Pleural lesions have been reported, but this is the first article describing PMP lesions in the thoracic spine, after dissemination to the thorax. This lesion caused vertebral pathological fracture and spinal cord compression at T6-T7 level, resulting in upper back pain and kyphosis as well as mild pyramidal syndrome, in a patient with a previously diagnosed PMP. A complete corpectomy of T6 and T7 was performed along with laminectomy, and posterior fixation involving three vertebrae above and below. Tumor resection was carried out to protect the spinal cord, without being "carcinologic".

The recommended treatment for PMP is a combination of cytoreductive surgery (CRS) with hyperthermic intra-peritoneal chemotherapy (HIPEC) associated with mitomycin-C and cisplatin (17). This treatment significantly improves patients' survival with respective rates of 77.8%, 79.5%, and 55.9% at 3, 5, and 10 years (18). However, there is significant morbidity associated with the use of combination treatment. Moreover, relapses can occur after this treatment, most commonly involving the peritoneum, but also pleural or systemic metastases in a relevant number of patients, who may require surgical resection of the recurrent lesion (19). Like abdominal lesions, intrapleural disseminations can be treated with hyperthermic intrathoracic chemotherapy (HITHOC) and pleural decortication (20). Some authors even

recommended hyperthermic intra-thoraco-abdominal chemotherapy to prevent pleural dissemination in case of diaphragm breach during CRS (21, 22). Hence, management of patients with PMP must be discussed in multidisciplinary meetings involving specialized healthcare providers such as oncologists, radiologists, and surgeons.

As it is the first description of PMP location in the spine, there is no recommendation to follow for the management of such lesions. Principles applied for surgical management of spinal metastases served as guidelines (23). In this case, the PMP lesion induced a triple spine-related issue: spinal cord compression, spinal mechanical instability, and sagittal malalignment due to major regional kyphosis. Hence, a circumferential spinal cord decompression was performed through laminectomy and corpectomy. Kyphosis was corrected by vertebral resection, and mechanical stability was insured by a large corpectomy cage in the front, and a +3/-3 pedicle screw construct posteriorly. As opposed to spinal metastases management with lower life expectancy patients, the maximum tumoral mass was resected to avoid local recurrence.

Vertebral resections are demanding techniques with high rates of complications, around 30% depending on the surgeon's experience (24). An alternative treatment could have been to perform a corpectomy by lateral approach through thoracotomy, thus achieving anterior decompression, and to complete the procedure with percutaneous pedicle screws. This option was ruled for two reasons: first, to avoid returning in the previously operated thorax, and second to perform a circumferential decompression as there were PMP lesions lateral to the spinal posterior arches.

Conclusion

PMP is a rare condition, with no previously described location in the spine. Here, a case of a PMP extension to the thorax causing a contiguous thoracic spine lesion and spinal cord compression was described. To achieve spinal cord decompression, kyphosis correction, and spine stabilization, a two-level vertebrectomy, laminectomy, and posterior fusion would be beneficial for a PMP lesion of the thoracic spine.

Conflict of Interest

The authors declare no conflict of interest in this study.

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