

A Huge Neurofibroma of the Lower Back Invading the Spinal Cavity

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A 56-year-old woman came to the hospital with a rapidly growing mass on her lower back (Fig. 1). Thirty years earlier, she had been diagnosed with neurofibromatosis type 1 (Von Recklinghausen's disease). The patient had multiple nodules, which were scattered over her entire body from her late twenties. The mass was palpable two months before



Fig. 1. Photograph of a 56-year-old female neurofibromatosis type 1 patient. A mass measuring 30×30 cm in size was found on the lower back.

presentation to our department, and it had been growing rapidly for the final three weeks. When lying down, the patient's performance of physical activity was impaired due to the mass. An abdominal-pelvic multi-detector computed tomography scan and sonogram showed a huge mass with a fluid-filled central portion located at the lower back. Increased peripheral vascularity and malignant transformation of the neurofibroma were suspected. In addition, a spina bifida-like lesion was noted at the spinous process of the L4 spine (Fig. 2). However, neurologic symptoms were not observed. Because there appeared to be a direct connection between the mass and the spinal nerve, additional imaging was required to distinguish the origin of the mass. On magnetic resonance images, the mass had no direct connection with the spinal canal. The mass measured 30×30 cm×18 cm in size and gave the impression of an abscess or a malignant schwannoma. Before the operation, aspiration was performed in order to reduce the size and to increase the mobility of the mass; 350 mL of bloody discharge was drained and spindle cell nests and nuclear atypia were observed on the cytological examination. Spindle cells were formed by abnormal mitosis or cell division, leading to the development of a cancerous tumor. The patient underwent wide excision under general anesthesia. A vertical spindle-shaped incision was made at the central portion of the mass. Dissection was performed between the skin flap and the peripheral muscle of the back. The skin was very thin but fast growing. Dissection was extended to the tendinous portion of the lower back muscle

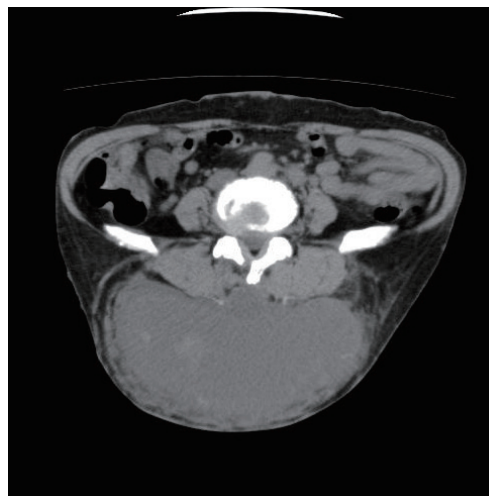


Fig. 2. Axial view of the abdomen on enhanced computed tomography scan shows a spina bifida-like appearance on the lumbar spine and fluid collection at the central portion of the mass.

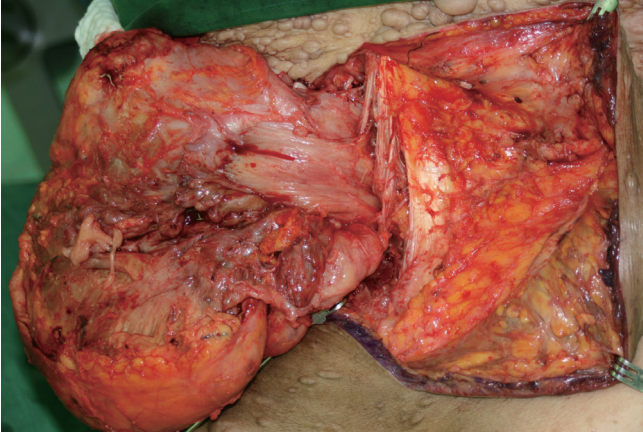


Fig. 3.

Photograph of the mass after dissection. The spinal cavity was perforated; however, no direct connection between the mass and the spinal nerve was observed.

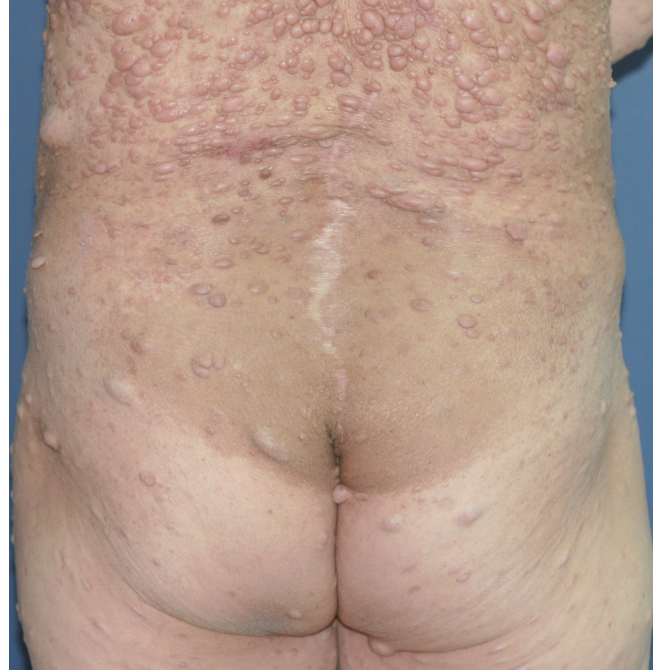


Fig. 5.

Photograph at postoperative two weeks. Skin contour was excellent, and there was no wound problem.

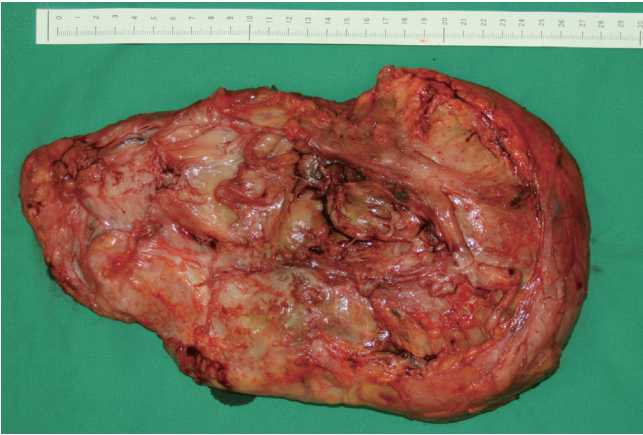


Fig. 4.

Photograph of an extracted mass after complete resection.

and spina bifida of the lumbar spine. On the base of the mass, dissection was performed between the lower back muscles and the mass. In the midsacral area, the mass was found to extend into the spinal cavity (Fig. 3); however, no direct connection with the spinal nerve was observed. After resection, the soft tissue defect was covered using a fasciocutaneous flap. The mass showed a well-encapsulated appearance with central necrosis. The mass was $28 \times 17 \times 7$ cm in size and weighed approximately 4 kg (Fig. 4). The external wound margin was invaded by a neurofibromatic change. However, given that the back was already entirely invaded by neurofibromas, the wound margin necessarily contained some neurofibromas. After two weeks, the patient recovered completely without any complications (Fig. 5). The mass was confirmed to be a type 1 neurofibroma, and

malignancy was not observed. The patient did not show any particular complaint for one year after the operation, and there has been no suspected sign of recurrence ever since. Neurofibroma is a benign tumor of the peripheral nervous system and could occur either accidentally or in association with neurofibromatosis type 1 (NF type 1). NF type 1 shows a high-frequency malignant transformation on peripheral nerve sheath tumors [1]. Most of the neurofibromas invade the skin and the superficial subcutaneous tissue. However, on various occasions, the mass can be located near major nerves and blood vessels, even intracranially [2]. Complete excision of the neurofibroma is challenging because of its infiltrating nature and tendency of recurrence [3]. Previous studies have shown that patients with NF type 1 are at a much greater risk of cancer, particularly leukemia, glioma, and malignant peripheral nerve sheath tumor (MPNST) [4]. Accordingly, NF 1 patients should undergo a regular checkup. On an aspiration exam, spindle cells were found. Because spindle cells have a potential for transformation into malignant cells, complete surgical resection is essential. Spindle cells are often found in cancerous tumors such as cellular schwannoma, synovial sarcoma, MPNST, dermatofibrosarcoma protuberans, leiomyosarcoma, clear cell sarcoma, and melanoma

[5]. We experienced a rare case of a huge neurofibroma suspected as a spina bifida on the lumbar level. As neurofibromas have a tendency toward malignant development, preoperative imaging is essential for distinguishing the character of the mass and the existence of metastasis. In addition, ongoing follow-up is critical for such cases.

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A Rare Case of Transitional Meningioma

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Meningiomas are a diverse set of tumors arising from the meninges, which are the membranous layers

surrounding the central nervous system. Most meningiomas grow inward toward the brain as discrete, well-defined, dural-based masses and are spherical or lobulated. The borders between the tumor and the brain are usually smooth and clear because they preserve the histologic structures, such as the tumor capsule, cerebrospinal fluid, arachnoid mater, and pia mater [1]. Considering such a growth pattern, we report a rare case of transitional meningioma misdiagnosed as a soft tissue tumor.

A 60-year-old female patient presented with a hard and fixed mass that grew spontaneously on her forehead within one year (Fig. 1). There was no history of trauma or exposure to carcinogens, and she was otherwise fit and well. Computed tomography (CT) showed frontal cortical thickening and swelling suggestive of a type of soft tissue tumor. To determine the character of the tumor-like mass, excision was performed (Fig. 2). The pathologist confirmed the mass as a transitional meningioma. Magnetic resonance imaging (MRI) which was taken after primary excision revealed an intracranial mass that was 3.0 cm in size, ovoid and well enhanced (Fig. 3). Wide-excision osteoplastic craniotomy *via* a bicoronal approach was performed under navigation guidance by neurosurgeon.

Transitional meningiomas are also called as mixed meningiomas, and these tumors have transitional between those of meningothelial and fibrous meningioma and they are common tumors with meningothelial, fibrous, psammomatous, and angioblastic meningiomas. 90% of meningiomas are consisting of those five types of meningiomas. The brain mass was also established as a meningothelial-type meningioma, 2.5 × 2.3 cm in size. Histologically, the tumor was lobulated by intersecting collagenous fibers. Immunohistochemical staining was positive for vimentin and epithelial membrane antigen and



Fig. 1. Preoperative photograph showing a lesion on the right forehead.