A Solitary Giant Neurofibroma of Inguinal Region—— A Case Report

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Case Report

Keywords: giant, neurofibroma, solitary, inguinal region, case report

Posted Date: September 22nd, 2020

DOI: <https://doi.org/10.21203/rs.3.rs-76761/v1>

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Version of Record: A version of this preprint was published at Science Progress on January 1st, 2021. See the published version at <https://doi.org/10.1177/00368504211004269>.

Abstract

Background

Neurofibroma is a rare nerve sheath tumor of neuroectodermal origin, especially the huge and isolated neurofibroma located in the inguinal region. To our knowledge, no such case has previously been reported.

Case presentation

We report a case of 34-year-old male patient with a 4-year history of progressive enlargement of the medial root mass in his left thigh with sitting and standing disorders along with pain. The tumor was completely removed by operation, and pathological diagnosis showed neurofibroma. There was no obvious neurologic defect after surgery, and no recurrence tendency was found in the follow-up of 2 years.

Conclusions

For a large solitary mass with slow growth and no malignant clinical manifestations for a long time, clinicians can not rule out the hypothetical diagnosis of neurofibroma, even though its growth site is very rare, such as this case of a huge tumor located in the groin. For neurogenic tumors, early operation should be performed, and the prognosis of patients after tumor resection is excellent.

Background

Neurofibroma is a benign nerve sheath tumor, which means it is neuroectodermal in origin(1), growing along a nerve or nervous tissue. It is extremely infrequent, accounting for only 5% of all benign soft tissue tumors, and may manifest as a slow-growing mass with benign clinical behavior over time(2). Therefore, it is a major point to shorten the time interval between symptom generation and definite diagnosis, and to operate early, which is related to the final prognosis of patients. The giant type of solitary neurofibroma without the presentation of type I neurofibromatosis(NF-1) is even rarer, we found literature about that in the head and neck(3, 4), retroperitoneal (5, 6), oral cavity(7), thigh(8), labia(9) and other uncommon parts. But this case describes a giant solitary neurofibroma located in the groin region, where no relevant case has been reported to the author's knowledge.

Case Presentation

The patient, 34 years old, was admitted to our hospital because of progressive enlargement of the tumor in his left inner thigh root for four years. He complained about sitting and standing disorders along with pain, and his daily life was inconvenient. But there was no pain, itching or local skin redness and ulceration in the protuberance area, and no fatigue, numbness or discomfort was found in the lower limbs. The patient had no previous history of trauma or NF-1, and denied the family history of Von

Recklinghausen's disease. Physical examination showed that the mass was about 10.0 × 9.0 cm in size, solid in quality, clear in boundary, and moderate in activity. Tinel sign in his inguinal region was positive. The muscle strength of lower limbs were normal, all deep and shallow reflexes were symmetrical, and no cafe-au-lait pigmentation or other signs of Von Recklinghausen's disease were found. Electromyogram suggested that the sensory nerve conduction velocity(SCV) of the affected side was slightly slower than the contralateral. There was no abnormalcy in laboratory tests. And X-ray film of the left lower limb in the positive and lateral position showed slight osteolytic changes in the ischial and pubic branches, the increased acetabular density with sharpened edges, and manifested a huge soft tissue shadow[Figure 1]. Magnetic resonance imaging (MRI) showed that there was a circular abnormal signal focus in the left pubic muscle space, about 8.2 × 7.4 × 8.4 cm in size with clear boundary, and had slightly lower signal on T1WI, high signal on T2WI. There was patchy or flocculent shadow of long T1 and short T2 signal in the mass with a visible capsule [Figure 2]. After injection of contrast medium, the capsule was significantly enhanced, while the inside of the mass was enhanced slightly and unevenly, and the boundary between the mass and the surrounding muscles was clear [Figure 3]. According to the course of the patient, the primary diagnosis was benign tumor and nerve source was considered first, so tissue diagnosis is recommended for surgical planning, while preoperative biopsy could be routinely and safely performed using ultrasound or CT guidance, but surgeons needed to pay attention to the risk of causing nerve or vascular damage, especially in this patient with a giant mass that might result in more narrow puncture gap and more fragile nerve fibers.

Then, under general anesthesia, the patient underwent radical resection of the tumor. After longitudinal myotomy of sartorius, it was found that the tumor with intact capsule originated from the femoral nerve, and the base of it was situated in the pubic muscle, some of the surrounding nerve fiber tracts were extruded extremely thin by it, and the whole mass was between the groin and the inner thigh [Figure 4]. Afterwards, a mass of 10.6 × 8.5 × 5.5 cm in size was dissected and excised, the femoral nerve was released, the operation field was cleared and a negative pressure drainage tube was placed, and then the tissue and muscle were stitched layer by layer, and the regional pressure applied. Pathological findings showed that the capsule was intact, the tumor cells were mainly spindle cells, the background was rich in mucus like substances, and the cells were in the shape of ticking tadpole [Figure 5]. The results of immunohistochemistry were CK (Pan) (-), Ki-67 (about 2% +), S-100 (+), desmin (-), CD34 (vascular+), SMA (-), β - Catenin (+), bcl-2 (-), CD99 (-) [Figure 6]. These proved the diagnosis of neurofibroma. After the operation, there was no neurologic defect in the patient, and he went to the outpatient clinic regularly every three months. Two years after follow-up, the daily life of patient had no adverse effect, and no local recurrence was discovered.

Discussion And Conclusions

Neurofibroma is a rare benign nerve sheath tumor composed of Schwann cells, perineural like cells and fibroblasts(10). It can be found in any peripheral nerve that can be invaded by neurofibroma, especially in lower limbs and upper limbs(2). There are generally three types: solitary, diffuse and plexiform. Among them, the solitary neurofibroma is usually a small polypoid mass with its maximum size of less than

2 cm(4, 11). Most neurofibromas appear as single lesions in dermis or subcutaneous, but there are still 10% cases related to NF-1, which is an autosomal dominant genetic disease, formerly known as Von Recklinghausen disease, with clinical features of cafe-au-lait pigmentation, benign skin and subcutaneous tumor known as neurofibroma, characteristic osteopathy, iris focal malformation(12, 13). In this case, we did not find a correlation performance. However, due to the effect of NF-1, this type of neurofibroma has larger volume, higher tendency of malignant transformation and higher recurrence rate (4, 8, 14). Therefore, when we encounter neurofibroma, we still need to actively look for other manifestations of Von Recklinghausen's disease to exclude the possibility of malignancy.

As the growth of neurofibroma is slow, and can occur at any age, has no sexual preference, and usually has no pain, numbness or other discomfort symptoms(15), its diagnosis is mainly depending on the result of imaging and microscopy. The differential diagnosis includes nodular fasciitis, myxoma, neurofibrosarcoma and adult rhabdomyoma, especially schwannoma(16, 17). Both schwannoma and neurofibroma belong to the peripheral nerve sheath tumor(7). They not only have homologous lesions, but also have similar imaging manifestations. Ultrasound can't distinguish them reliably, for their sonographic features are medium echo solid extension mass, generally without blood flow signal, which are the common features of soft tissue tumors(2). In the X-ray manifestations, since its soft tissue contrast is worse than that of MRI, and the bone is clearly displayed, X-ray is often used to judge the bone invasion or destruction of peripheral nerve tumors (30% – 50% of patients can see), such as aggressive damage, bone compression and displacement, external cystic change of bone, subperiosteal cyst and pseudarthrosis of tibia, etc.(18), which is also presented in our case, specifically the osteolytic manifestations of the ischial and pubic branches. But these imaging feature contribute a little to the differential diagnosis. Unfortunately, the MRI, which is the most sensitive imaging method for soft tissue, manifests neurofibroma may similar to those of schwannoma. They usually show low or equal signal compared with muscle on T_1 -weighted image(T_1WI), high signal on T_2 -weighted image(T_2WI), and can change with the enhancement of contrast agent, especially when the tumor has mucoid mechanism, it can show no or slight enhancement(8, 15). It is worth noting that compared with schwannoma, neurofibroma often lacks complete capsule, which may help us to distinguish common cases by MRI images(6). At the same time, when the myxoid tissue in the neurofibroma is rich enough, it can show a characteristic image from the schwannoma, which presents the low T_2 signal central area of collagen tissue as the bull's eye, and the high T_2 signal of the myxoid tissue as peripheral area(8). However, this case is extremely rare. The solitary mass is not only huge in size, particular in tumor site, but also has a complete capsule on MRI without the target sign, which leads to our erroneous initial diagnosis of schwannoma.

The final diagnosis depends on the pathological examination of the tumor, which is done by detecting the nerve cells contained in the tumor, as shown in this case. First, unlike schwannomas, the parent nerve of neurofibromas is usually separable in plexiforms rather than isolated ones(6). Secondly, neurofibroma is rich in mucoid tissue (Antoni B area), while schwannoma has a compact stroma (Antoni A area)(8). In addition, although both of them are composed of fusiform nerve sheath cells, the nuclei of neurofibroma

are slender and wavy, while the nuclei of schwannoma are oblong and arranged closely and parallel to each other in palisade or incomplete whorls (Verocay bodies)(4, 15). Immunohistochemical results also provide the basis for differentiation, which is reflected in the uneven staining of S-100 protein, only local immune response to epithelial membrane antigen (EMA), and strong CD34 positive in neurofibroma, while schwannoma cells showed more intense and uniform staining of S-100 protein, and the capsule stained EMA(2, 4).

When clinical diagnosis of neurogenic tumor is made, we should routinely and safely performed biopsy using ultrasound or CT guidance to achieve pathological diagnosis, along with the tissue diagnosis recommended for surgical planning. But biopsy has a risk of causing nerve damage, which is likely to occur in this case as intraoperatively, we found that some of the nerve fiber tracts were extruded particularly thinly by the tumor. At present, the first choice of clinical treatment for neurofibromas is still complete resection, especially when symptoms occur or appearance are affected, the postoperative prognosis is good and the recurrence rate is low(2, 10). However, the parent nerve of neurofibromas is usually unseparable in solitary neurofibromas(6). Thus, it should be noted that under the premise of complete resection, the scope of tumor resection should be precisely determined, and microsurgery is recommended, at the same time, the parent nerve and non-involved nerve fibers need to be preserved as much as possible, so as to avoid serious risk of nerve injury and the sacrifice of the parent nerve. These require microsurgical skills and rich surgical experience of the chief surgeon. Alternative therapies include enucleation and subtotal resection, which are often used in the patients that completely resection may cause severe functional defects or cosmetic deformities. However, due to the inevitable recurrence caused by the residual tumor tissue, they are often not considered(6). Radiotherapy is also a feasible method, but it is only suitable for isolated lesions to control or reduce the growth of lesions, and it is effective for the treatment of juvenile angiofibroma(19). Recently, there is also a literature showing that photodynamic therapy is effective for solitary neurofibroma, but its long-term survival rate and long-term prognosis still need to be investigated(10).

In consideration of the limitation and huge type of the focus in this case, as well as the adverse effects on daily life, such as sitting and standing posture change obstacles and pain, we performed a complete resection of the tumor in this patient. After incision of the outer membrane, we peeled off the tumor, kept the normal nerve fiber bundle as much as possible, protected the large blood vessels, and covered the nerve blood vessel bundle with the re-suture of the muscle after resection. There was no obvious discomfort in the patient after operation, and no daily life disorder and tumor recurrence were found in 2-year follow-up.

In general, this paper presents a rare case of giant solitary neurofibroma located in the groin area without NF-1 performance. It is a benign developing peripheral nerve tumor, and its pathogenesis is still unclear. Although the majority of neurofibromas have no discomfort symptoms at early stage, as they grow slowly, when they are too large or too deep, they can cause neurological symptoms or compression symptoms of adjacent organs just as what had happened in this patient, such as Tinel sign positive, and may also bring difficulty to operation and cause poor prognosis. Therefore, early diagnosis should be

carried out to shorten the time interval between symptom appearance and treatment, which can be achieved by MRI and clinical manifestations. The confirmed diagnosis is through pathological sections. Except for the plexiform type, most of the tumors show good limitations, so the gold standard for the treatment of this disease is still surgery, especially in this case of large and isolated neurofibroma. Good prognosis and low recurrence rate are obtained after radical resection.

Abbreviations

Sensory nerve conduction velocity SCV; Magnetic resonance imaging MRI; type I neurofibromatosis NF-1; T₁-weighted image T1WI; T₂-weighted image T2WI; epithelial membrane antigen EMA.

Declarations

Ethics approval and consent to participate

The study protocols were approved by the Medical Ethics Committee of the First Affiliated Hospital of the College of Medicine, Zhejiang University

Consent for publication

Written informed consent was obtained from the patient for publication of clinical details and clinical images. Upon request, a copy of the consent form is available for review by the Editor of this journal

Availability of data and materials section

The dataset supporting the conclusions of this article is included with the article.

Competing interests

The author declares that they have no competing interests.

Funding

The study was funded by the National Natural Science Foundation of China (the grant number 81702135), Zhejiang Provincial Natural Science Foundation (LY20H060007), the Zhejiang Traditional Chinese Medicine Research Program (grant number 2016ZA124, 2017ZB057) and Zhejiang Medicine and Hygiene Research Program (grant number 2016KYB101, 2015KYA100). The funding bodies had no role in the design of the study; in collection, analysis, and interpretation of data; and in drafting the manuscript.

Author contributions

HL designed the study, performed data collection, analyzed the results, and HY Z drafted the manuscript. All the authors have read and approved the final manuscript.

Acknowledgements

Not Applicable

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Figures

Figure 1

In the anteroposterior(A) and lateral(B) radiographs of X-ray, there were slight osteolytic features of ischial and pubic branches, along with the increased acetabular density and sharpened edges, and manifested a huge soft tissue shadow which was indicated by the red arrows.

Figure 2

In the anteroposterior(A) and lateral(B) radiographs of X-ray, there were slight osteolytic features of ischial and pubic branches, along with the increased acetabular density and sharpened edges, and manifested a huge soft tissue shadow which was indicated by the red arrows.

Figure 3

After injection of contrast medium, the capsule was significantly enhanced, while the inside of the mass was enhanced slightly and unevenly, and the boundary between the mass and the surrounding muscles was clear.

Figure 4

After muscle incision of sartorius, it was found that the tumor with intact capsule originated from the femoral nerve, and the base of it was located in the pubic muscle, the whole mass was between the groin and the inner thigh, about 10.6X8.5X5.5cm in size.

Figure 5

Pathological findings (HE stained): the capsule was intact (A) X 50, the tumor cells were mainly spindle cells (B) X400, the background was rich in mucus like substances (C) X400, and the cells were in the shape of ticking tadpole (B).

Figure 6

The results of immunohistochemistry(X400): (a),CK (Pan) (-); (b), Ki-67 (about 2% +); (c), S-100 (+); (d), desmin (-); (e), CD34 (vascular+); (f), SMA (-); (g), β - Catenin (+); (h), bcl-2 (-);(i), CD99 (-).

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