Andrology and fertility

A case report of solitary neurofibroma of the Vas deferens

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ABSTRACT

Solitary neurofibroma of the male genital tract is a rare benign peripheral nerve sheath tumor which is considered to originate from the Schwann cell. Neurofibroma of the male genital tract has been reported extremely rarely in the literature. We present a case of neurofibroma of the Vas deferens in a 59-year-old patient. The clinical and pathological data of a patient with Neurofibroma of the Vas deferens were retrospectively analyzed and radical resection of the left side spermatic cord tumor was performed. This case report will help in understanding this rare tumor.

Introduction

Solitary neurofibroma of the male genital tract is composed of Schwann cells and fibroblasts. Although it is a benign lesion with very low malignant potential, especially in the absence of neurofibromatosis type 1 (NF1), accurate diagnosis is still important because it may lead to serious complications and psychological distress. Isolated intrascrotal neurofibroma unrelated to NF1 is very rare. According to the literature review, only 14 cases are reported.1 Here we report a confirmed case of neurofibroma of vas deferens.

Case report

A 59-year-old man, with no significant past medical history, was admitted to our hospital because of the accidental discovery of mass in the upper left scrotum 2 years ago. There was no pain and other symptoms of the genitourinary system. Physical examination showed a mass in the spermatic cord above the right scrotum near the outer inguinal ring with no tenderness, medium quality, smooth surface, good mobility, and no adhesion with scrotal skin. No abnormalities were found in bilateral tests, epididymis and right spermatic cord. Ultrasonography showed that the left scrotum had a low echo mass of about 38 mm × 20 mm below the spermatic cord, with clear boundary, irregular shape, uniform internal echo and abundant blood flow signals in color Doppler flow imaging (CDFI) (Fig. 1a). No enlarged lymph nodes were found in the inguinal, pelvis and retroperitoneum. The levels of serum AFP, beta-HCG and lactate dehydrogenase (LDH) were all within the normal range.

During the surgery, we found that the mass was located on the internal spermatic duct above the left testis (Fig. 1b). The mass was encapsulated, with a clear boundary and smooth surface (Fig. 1c). Histopathology showed that the specimen was 3 cm × 2.2 cm × 2 cm in size, the surface was gray-white, smooth, with a complete capsule, and the profile was substantially fish-like and gray-pink. On Light microscopic, HE staining the lesion was a typical neurofibroma consisting of bundles of spindle cells and dark wavy nuclei separated by collagen chains (Fig. 2). Mast cells and focal hemosiderin were also seen. Immunohistochemical staining were: Vim (+), CD34 (Part +), S100 (+), Stat-6 (--), Caldesmon (Part-), DES (--), SMA (--), Bel-2 (--), GFAP (--), catenin (--), Ki67 (+3%) (Fig. 3a and b). The pathological diagnosis was neurofibroma of the left vas deferens. There was no recurrence or metastasis after a 1-year follow-up.

Discussion

Neurofibroma is a benign peripheral nerve sheath tumor, composed of Schwann cells and fibroblasts. These tumors are rich in collagen fibers and, contrary to schwannomas, they are not enclosed by nerve sheaths and can infiltrate between nerve bundles. The Immunohistochemical study showed that neurofibroma expressed S-100 protein. There are three types of neurofibroma: localized, diffuse and plexiform. Among the 14 cases, reported in the literature,9 cases are spermatic neurofibroma and 5 are intrascrotal. This case was localized neurofibroma.

The exact origin of scrotal neurofibroma is unclear. Some authors believe that it originates from the reproductive branch of the genitofemoral nerve located behind the spermatic cord.2 Some authors suggest

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that solitary neurofibroma attached to spermatic cord may originate from ilioinguinal nerve or even smaller axons. Evidences show that the main nerve reflex pathways controlling vas deferens are lumbar sympathetic chain, lumbar visceral nerve, epigastric hypogastric plexus, hypogastric nerve and, pelvic plexus. The latest development of neuro-markers (S-100 protein, protein gene product 9.5, etc.) showed that in the connective tissue of the adventitia of the vas deferens, branches along the nerve trunk extending along the vas deferens to the adventitia of the vas deferens, and then branches repeatedly to form dense intramuscular nerve plexus and further to form nerve plexus at the base of the inner epithelium. The reproductive branches of the genital femoral nerve originating from these tumors were usually located on the back of the spermatic cord, supplying the levator muscle, and then distributed to the scrotum and the skin adjacent to the thigh. Considering that, the tumors in this patient did not attach to the testis, epididymis or spermatic cord, but extended and infiltrated along the vas deferens. We believe that the neurofibroma in this patient is more likely to originate from the pelvic plexus, the epididymal branch (testicular nerve), rather than from the genitofemoral nerve of the lumbar plexus.

Neurofibroma of the spermatic cord is characterized by palpable masses, dull pain or loss of sensation. From the literature review, only 2 of the 14 reported cases showed pain. This case is also having no pain. Ultrasonography of solitary neurofibroma usually showed a well-defined, relatively homogeneous, hypoechoic mass, and lacks to moderate blood flow on color Doppler imaging. Ultrasonography of this case showed a low echo mass with clear boundary, irregular shape, uniform internal echo, and abundant blood flow signals in color Doppler flow imaging (CDFI). MRI is the preferred method for the characteristics and diagnosis of neurofibroma. MRI can provide accurate anatomical analysis of the lesion and its relationship with surrounding soft tissues. No MRI was performed in this case. Intrascrotal neurofibroma should be differentiated from the scrotal mass of paratesticular origin, including spermatic cord, epididymis, embryonic residue and sheath origins, such as leiomyoma, fibroma, lipoma, and hemangioma.

Surgical resection is the choice of treatment for solitary neurofibroma unrelated to NF1. Intraoperative frozen section diagnosis is useful in excluding malignant tumors and avoiding orchiectomy. However, they are not encapsulated and infiltrate between nerve bundles, maternal nerves must be sacrificed, therefore conservative treatment or incomplete resection is preferred to minimize nerve and permanent injury. In this case, the tumors grew and infiltrated along the vas deferens, and consequently, partial vasectomy was performed. Solitary neurofibroma usually has a good prognosis. Malignant transformation is 4% in NF1 patients and very rare in the absence of NF1. Local recurrence is also uncommon in non-NF1-related lesions.

**Conclusion**

Neurofibroma of the Vas deferens is an extremely rare benign tumor. Preoperative diagnosis is difficult. Surgical resection is the treatment of
choice and the final diagnosis is provided by histopathology.

Consent

Written informed consent was obtained from the patient for publication of this case report.

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Declaration of competing interest

None of the contributing authors has any conflict of interest.

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References

1. Gupta S, Gupta R, Singh S, Pant L. Solitary intrascrotal neurofibroma: A case diagnosed on aspiration cytology. Diagn Cytopathol. 2011;39(11):843–846.
2. Gupta S, Ruchika Gupta R, Singh S, Pant L. Solitary intrascrotal neurofibroma: A case diagnosed on aspiration cytology. Diagn Cytopathol. 2010;39(11):843–846.
3. Hosseini MM, Geramizadeh B, Shakeri S, et al. Intrascrotal solitary neurofibroma: a case report and review of the literature. Urol Ann. 2012;4(2):119–121.
4. Milathianakisa KN, Karamanolakisa DK, Mpogdanaona IM, Trihia-Spyrou EI. Solitary neurofibroma of the spermatic cord. Urol Int. 2004;72(3):271–274.
5. Johnson DE, Kaesler KE, Mackay BM, Ayala AG. Neurofibrosarcoma of spermatic cord. Urology. 1975;5(5):680–683.