Urology Case Reports 6 (2016) 21–23

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: www.elsevier.com/locate/eucr

Oncology

Latent Progression Pediatric Scrotal Schwannoma. A Case Report

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ARTICLE INFO

Article history:
Received 5 December 2015
Received in revised form
27 December 2015
Accepted 29 December 2015

Keywords:
Paediatric schwannoma
Scrotal

ABSTRACT

We report the case of a 24 year old patient being diagnosed with scrotal schwannoma initially presenting at age 9. To our knowledge, this is the first case with such an early onset. The patient underwent an uncomplicated surgical excision. We are also reviewing the literature on scrotal schwannoma.
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Introduction

Schwannomas are mostly benign nerve tumors that arise from Schwann cells. The incidence of schwannomas is very rare and they are equally presented in both sexes. The most common regions of the human body that schwannomas develop are head and neck.[1] *Acoustic neuroma* is the most common type of benign schwannoma, which can cause deafness. The presentation may be spontaneous or part of a syndrome that is called neurofibromatosis or schwannomatosis.[2]

In the literature, cytological features of the tumor are well documented. Although extremely rare, cases of scrotal origin of such tumors have been reported. Here we report a case of intrascrotal extratesticular solitary schwannoma in a 24 year-old patient. The nodule initially presented at age 9. To our knowledge, it is the first case of scrotal schwannoma in a patient of such a young age presented in literature.

Our case

A 24 year-old gentleman was referred by his GP for a left side inferior solitary scrotal nodule. He initially noticed its presence about 15 years ago. Although the patient has always been completely asymptomatic, the nodule became intermittently tender and painful in the last couple of years. The patient was fit and his medical history was clear. His family history was clear for neurofibromatosis or schwannomatosis.

Clinical examination showed a superficial, well defined, mobile, small-sized nodule. The lesion was distinct from the left testis. Both testes and epididymi were normal and no palpable lymph nodes were noted in physical examination.

Urgent scrotal ultrasound scan showed a 8.7 × 7 × 5.6 mm well defined solid mass in the left posterior hemiscrotum, closely adjacent to but not inside the left epididymal head. It casted posterior enhancement and there was internal blood flow noted. No other scrotal pathology was noted (Fig. 1).

Full blood count, urea and electrolytes readings were within normal range. Testicular tumor markers were within normal range.

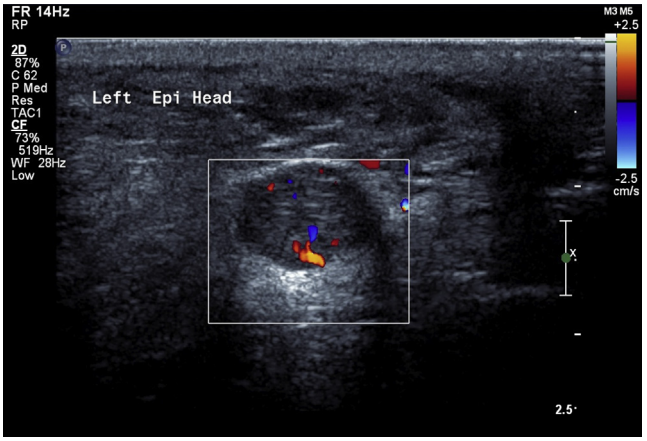

Figure 1. Doppler sonographic image of the scrotal nodule.

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http://dx.doi.org/10.1016/j.eucr.2015.12.012

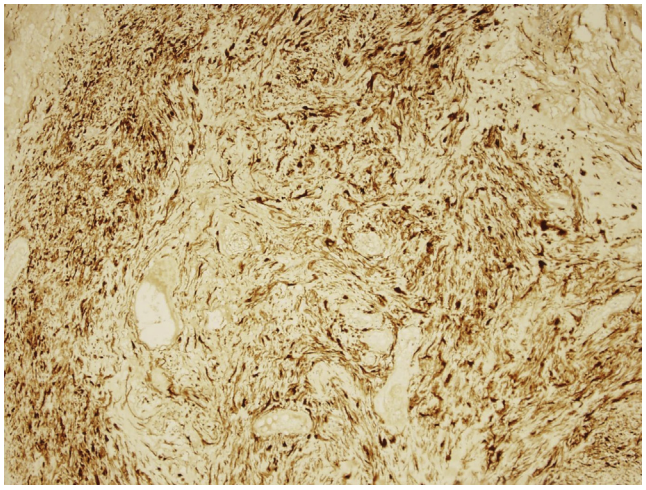

Figure 2. Schwannoma S-100 immunohistological staining image (×200).

The case was discussed at our local Multi-Disciplinary Team meeting (MDT) where excision of the nodule was recommended. The patient was fully informed and consented for the above procedure.

Intraoperatively, the left testis was explored through a mid-scrotal incision. The mass was completely excised and tunica vaginalis eversion was carried out.

Macroscopic evaluation of the specimen revealed a 9 mm translucent nodule. The histopathology report showed a well circumscribed and encapsulated schwannoma. Immunostain for s-100 was strongly positive and Epithelial Membrane Antigen staining (EMA) was negative (Fig. 2). Surgical margins were clear.

Discussion

Schwannomas originate from Schwann cells that are responsible for the production of myelin, which is a fundamental substance of the sheath of the peripheral nerves. Their incidence is not specified because of their rare occurrence. They are more common between the second and forth decade of life and are equally presented in both sexes. Most cases occur in the neck and head region (trigeminal, facial, vestibular and vagus nerve, the parotid and thyroid gland, vocal cords, floor of the mouth, orbit, and infra-temporal fossa).[2] Other less common locations include the extremities, mediastinum, thorax, retroperitoneum, pancreas and pelvis. Their presentation at perineum and scrotum is very rare. There are ten (10) reports of schwannomas of the scrotum in the literature, one of the lesser pelvis, one of the prostate and two of the penis (as noted in Table 1).

The clinical findings of male genital schwannomas are non-specific. Usually these tumors are asymptomatic until they grow enough to create pressure symptoms. Those could involve hematuria, lower abdominal discomfort and mild to moderate dysuria depending on the site of presentation.

Table 1
Schwannoma review table.

Author	Pt age	Sight of presentation	Solitary/ multiple	Pre-op Examinations	Management	Histology Results	Follow up
1) Young-Joo Kim et al.[3]	67	Intrascrotal/extratesticular	Multiple	Testicular tumor markers, scrotal USS	Complete excision	S-100 positive, SMA negative	6 years, no recurrence
2) Sebastiano Pante et al.[4]	64	Solid pelvic mass compressing corpus spongiosum of membranous urethra and left corpus cavernosum	Solitary	USS, CT, MRI, retrograde urethroscopy, echo-guided biopsies	Complete excision through arcuate incision in the urogenital triangle.	Antony A&B growth patterns, S-100 positive, “ancient schwannoma”	4 years, no recurrence
3) Mohammad Shahid et al.[5]	45	Root of the scrotum	Solitary	Testicular markers, USS	Complete excision with partial scrotectomy	Antony A&B growth patterns, Verocay bodies	No f/u results
4) Michelle Bergeron et al.[6]	16	Intrascrotal, root of penis	Solitary	Testicular tumor markers, USS	Complete excision	S-100 positive	N/A
5) Giovanni Palleschi et al.[7]	52	Intrascrotal/extratesticular	Solitary	Testicular tumor markers, USS with doppler and elastography	Complete excision	Antony B pattern, S-100/ vimentin positive	No f/u results
6) Peter Chan et al.[8]	28	Intrascrotal/extratesticular	Solitary	USS, CT, diagnostic wedge biopsy,	Complete excision	S-100 positive, “ancient schwannoma”	No f/u results
7) Ryo Ikari et al.[9]	66	Intrascrotal/extratesticular-head	Multiple	USS, MRI	Complete excision	Antony A&B growth patterns	No f/u results
8) V. Bhanvadia et al.[10]	32	Intrascrotal/extratesticular, ulcer of the scrotal skin over the lesion	Solitary	USS, FNA	Complete excision	Antony A&B growth patterns	N/A
9) Rui Jiang et al.[11]	38	Intrasctotal/extratesticular	Solitary	Testicular tumor markers, USS, frozen sections	Complete excision	Encapsulated, 20 mitosis per 10 high-power fields, malignant schwannoma, S-100 weakly positive	Metastatic disease, death 1 year after operation
Rui Jiang et al.[11]	65	Intraprostatic	solitary	FR, Flexible cystoscopy, USS, CTU	Transvesical suprapubic prostatectomy	Antony A pattern, S-100 positive	2 years, no recurrence
Rui Jiang et al.[11]	20	Left scrotal skin, adherent to the cord	Solitary	Testicular tumor markers, USS	Complete excision	S-100 positive, SMA negative	6 years, no recurrence
Rui Jiang et al.[11]	28	Coronal sulcus of the penis	Solitary	Biopsy	Partial penectomy	S-100 positive, SMA negative	6 years, no recurrence
Rui Jiang et al.[11]	34	Mid third of the penile shaft	Solitary	N/A	Complete excision	S-100/vimentin positive	4 years, no recurrence
10) Tarek Salem et al.[12]	45	Intrascrotal/extratesticular	Solitary	USS	Complete excision	Antony A&B growth patterns	3 months, no recurrence

CT, Computed Tomography; CTU, Computed Tomography Urogram; FNA, Fine Needle Aspiration; MRI, Magnetic Resonance Imaging; USS, Ultrasound Scan.

In previously reported cases, ultrasound scan, Computed Tomography and an MRI scan have been used to reach a more exact diagnosis. Tissue architecture is required for diagnosis but cannot be obtained from the cytology specimen.[8] Chan et al, performed wedge biopsy before tumor excision from the scrotum.[8]

Surgical excision remains the mainstay of treatment.

The microscopic appearance of schwannoma is distinctive, with two recognizable patterns. Antoni A areas are composed of compacted spindle cells often arranged in palisades or in an organoid arrangement (Verocay bodies).[8] Antoni B areas consist of tumor cells suspended in a myxomatous matrix that may appear microcystic. Positive immunostaining for the s-100 protein coupled with a negative reaction to CD34 and smooth-muscle actin and desmin are required to confirm the diagnosis of schwannoma.

The expression of high concentrations of S-100 protein in the cytoplasm of tumor spindle cells on immunostaining is useful in the differentiation of a benign schwannoma from a malignant peripheral nerve sheath tumor and from other benign spindle cell tumors. Malignant transformation of those tumors have a poor prognosis.

Most authors suggest follow up even in benign cases. Further investigation is recommended for patients with multiple schwannomas.

Multiple schwannomas in the same individual suggest two types of underlying tumor predisposition syndromes: neurofibromatosis type 2 and schwannomatosis. Both of these diseases include familial and non-familial cases. Patients with schwannomatosis are at increased risk for developing new schwannomas anywhere in the body although the long-term outcome of schwannomatosis is favorable.[9]

Conclusion

Our case is a presentation of a solitary scrotal schwannoma with an onset on a very young age and very slow progression. Although rare, schwannoma histological features are well established and surgical excision is the mainstay of treatment. Further investigation is needed in every single case to exclude correlation with neurofribomatosis.

Consent

The patient has given his consent for the images of his examinations to be published. A copy of the written consent is available, at anytime, for review by the Editor of this journal.

Conflict of interest

None.

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