

Case Reports

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TESTICULAR ALBUGINEA NEUROFIBROMA: FINDINGS AT ULTRASONOGRAPHY AND MAGNETIC RESONANCE IMAGING WITH PATHOLOGICAL CORRELATION

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Summary.- OBJECTIVES: To report the second case of solitary neurofibroma arising from the tunica albuginea in the literature and to show its imaging findings.

METHODS/RESULTS: We present a case of neurofibroma arising from the tunica albuginea in an adult patient not affected by neurofibromatosis. We describe the ultrasonographic and magnetic resonance imaging (MRI) features

and the histopathological characteristics along with a brief bibliographic review.

CONCLUSION: MRI may be useful to characterize paratesticular lesions. Neurofibroma should be included in the differential diagnosis when MRI depicts a well-circumscribed tumour with high-signal intensity on T2 and marked enhancement after gadolinium administration.

Keywords: Neurofibroma. Magnetic resonance imaging. Ultrasonography. Paratesticular pathology. Scrotum.

Resumen.- OBJETIVO: Comunicar el segundo caso descrito en la literatura de neurofibroma solitario con origen en la túnica albugínea testicular y los hallazgos radiológicos.

MÉTODO/RESULTADOS: Presentamos un neurofibroma de la túnica albugínea en un paciente adulto no afectado por neurofibromatosis. Se describen los hallazgos de este tumor en ecografía-doppler y resonancia magnética (RM) con correlación histopatológica, así como una breve revisión de la literatura.

CONCLUSIÓN: La RM puede ser una técnica útil para ayudar en la caracterización prequirúrgica de las lesiones paratesticulares. El neurofibroma debe ser incluido en el diagnóstico diferencial de masas escrotales cuando están bien delimitadas y muestran hiperintensidad de señal en T2 y realce intenso tras la administración de gadolinio.

Palabras clave: Neurofibroma. Resonancia magnética. Ecografía. Patología paratesticular. Escroto.

INTRODUCTION

Solid paratesticular tumors are rare lesions that often pose a diagnostic dilemma for both urologists and radiologists due to their rarity, the difficulties in distinguishing them from intratesticular lesions on physical examination, and ultrasonography (US) (1). Most of them are benign lesions, usually comprising lipoma, adenomatoid tumor, leiomyoma, hemangioma, and fibroma. Malignant lesions include liposarcoma, fibrosarcoma, rhabdomyosarcoma, lymphoma, and metastases.

The solitary neurofibroma is a benign neoplasm of the nerve sheath arising from the schwann cell, not related to neurofibromatosis, that has rarely been described in the external genitalia. There are about 11 cases of intrascrotal neurofibroma reported (2-12), and only one of them arised from the tunica albuginea (2). The magnetic resonance imaging (MRI) features of the intrascrotal neurofibroma have not been previously described. We present a case of unsuspected paratesticular neurofibroma and describe the ultrasonographic and MRI findings with pathological correlation.

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CASE REPORT

A 60-year-old-man presented with discomfort in the left inguinal area and scrotum 15 months after surgical left inguinal hernia repair. Physical examination revealed a normal-appearing surgical scar and a non-tender 1 cm mass at the lower pole of the right testis. There were no clinical signs of neurofibromatosis. Tumour markers were negative. Colour-doppler US of the right scrotum showed a 10 mm well-defined solid vascularised capsulated lesion. It bulged slightly distorting the testicle contour and was considered to be intratesticular. MRI demonstrated a right scrotal lesion hypointense and markedly hyperintense to testicular parenchyma on T1 and T2-weighted images (WI) respectively, demarcated by a fine low-intensity band on both sequences. After intravenous administration of gadolinium it enhanced strongly with a heterogeneous central area.

A right orchiectomy was performed. Macroscopic examination revealed a 10-mm well-circumscribed gelatinous extratesticular mass adherent to the tunica albuginea of the inferior pole of the right testicle. Histologically, the lesion consisted of bundles of spindle cells surrounded by abundant myxoid stroma with a rich capillary network. On immunohistochemical studies the neoplastic cells stained for S-100 protein and they were negative for CD31 and muscle-actin. They did not exhibit atypia and the proliferative index was very low. Neurofibroma was diagnosed based on these immunohistochemical data.

The postoperative course was uneventful and the patient remains free of relapse 18 months after surgery.

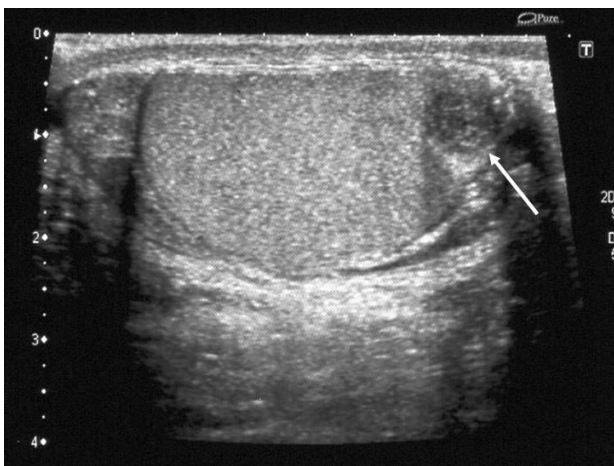


FIGURE 1. Longitudinal ultrasound image of the right hemiscrotum shows an approximately 1 cm well-defined lesion (arrow) with heterogeneous echotexture at the lower pole of the testis. The tumour bulges the contour of the testicle, being difficult to precisely determine its anatomic location.

DISCUSSION

The most common lesions of the testicular tunica are reactive disorders such as fibrous pseudotumour, followed by benign neoplasms (adenomatoid tumour), being malignant tumours less frequent. Other rare neoplasms include mesothelial tumours with müllerian differentiation and ovarian-type epithelium, Brenner tumour, hemangioma, spindle cell tumours (fibroma, fibrolipoma, leiomioma, malignant fibrous histiocytomas, leiomyosarcoma).

Neurofibroma is a benign neoplasm of the nerve sheath arising from the schwann cell. Neurofibroma can be solitary or multiple and has been associated with neurofibromatosis type 1 in some cases (4, 8). Location in the scrotum is extremely rare (2-10). There seems to be different anatomic sites of origin of the intrascrotal neurofibroma in the cases reported including the testis (3), tunica albuginea (2), subcutaneous scrotal tissue (4, 9), the genital branch of genitofemoral nerve (6), spermatic cord (7, 11, 12), and, occasionally, the exact origin could not be clearly identified (8, 10). Age at presentation is variable, ranging from 8 to 77 years. Most patients present with an asymptomatic slowly growing mass or unspecific local discomfort. In our patient neurofibroma was probably a coincidental finding on US performed to evaluate nonspecific contralateral discomfort.

US is the primary imaging technique for scrotal lesions assessment (1). However there are limitations. The findings of solid paratesticular tumors on US are often nonspecific precluding a presurgical diagnosis. In addition, it can be difficult to determine the origin of lesions originating on the capsule or very firm extratesticular tu-

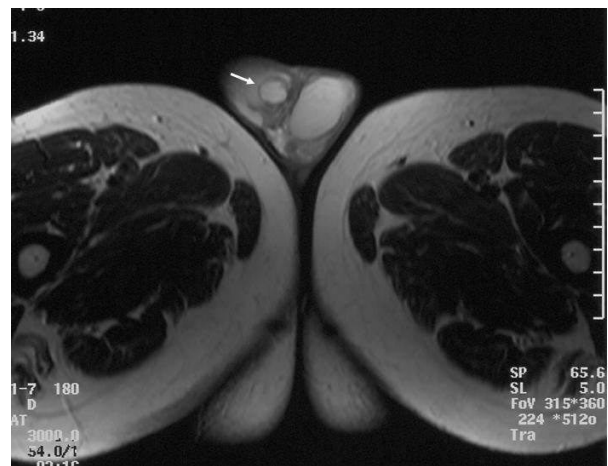


FIGURE 2. Transverse T2 weighted image shows the tumour (arrow) demarcated from the adjacent testicle by a low-signal band corresponding to a capsule or the tunica albuginea. Note the high signal intensity of the mass.

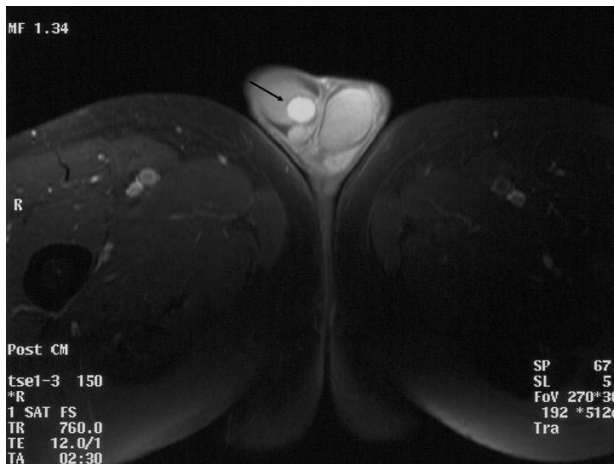


FIGURE 3. Transverse gadolinium-enhanced fat-saturated gradient-echo T1 weighted image demonstrates enhancement of the tumour (arrow) higher than that of the testicular parenchyma.

mors indenting the capsule and protruding into the testis (1). In our case, US depicted a solid vascularized lesion with a heterogeneous echostructure. Türkyılmaz et al (8) also found the intrascrotal neurofibroma to present as a heterogeneous solid mass, whereas Deliveliotis et al (7) described it as a hyperechogenic lesion.

MRI can be useful to help localization and characterization of extratesticular masses in selected cases. MRI allows confident diagnosis of epididymal cysts which are depicted as well-demarcated lesions with low signal intensity on T1-WI, high signal intensity on T2-WI, without enhancement after gadolinium intravenous administration. Lipoma typically is bright on T1 and hypointense on fat-suppressed T1-WI. Fibrous pseudotumor appears as an unenhancing lesion with low signal intensity on both T1 and T2-WI owing to fibrosis. Adenomatoid tumor appears as a mass slightly hypointense to testicular parenchyma on T1 and T2-WI that enhances less than normal testis (1). In our case, the neurofibroma appeared as a well-margined mass surrounded by a band of decreased signal intensity which corresponded on the specimen with the tunica albuginea, therefore, indicating the extratesticular origin. The lesion was hypointense on T1 and markedly hyperintense on T2-WI and showed intense enhancement. The myxoid stroma and the rich capillary network are probably responsible for the high signal intensity on T2-WI and the striking contrast respectively.

CONCLUSION

MRI may be useful to characterize paratesticular tumours in selected cases and narrow the differential diagnosis when clinical and sonographic features are inconclusive. Neurofibroma should be included in the differential diagnosis of such lesions when MRI depicts a well-circumscribed tumour showing hyperintensity on T2 and

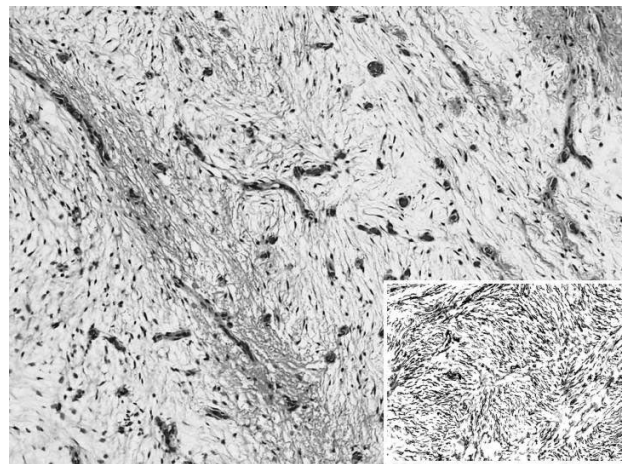


FIGURE 4. Photomicrograph shows spindle cells surrounded by abundant myxoid stroma and small capillary vessels (original magnification $\times 200$; hematoxylin-eosin stain). Inset photograph: neoplastic cells show S-100 positivity (original magnification $\times 200$).

marked enhancement after gadolinium administration, which are unusual findings for the vast majority of paratesticular lesions. The final diagnosis of neurofibroma, however remains immunohistopathological, being the positivity for S-100 protein key to the diagnosis.

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