Lymphoma of the scrotum in patients with Down's syndrome: US appearance. Mini-pictorial essay

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KEYWORDS

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Abstract Down's syndrome is relatively common, and patients who are affected have an increased risk of developing acute leukemia, but not solid tumors. Studies performed in larger patient populations have shown that solid tumors, including lymphomas, are significantly less frequent in Down patients than in children and adults who are not Trisomy 21-affected. Testicular lymphomas are rare and extremely aggressive. Ultrasound (US) combined with color Doppler is essential in the diagnosis and evaluation of treatment results in these lesions. As they are very rare, it was decided to publish this mini-pictorial essay.

Sommario La sindrome di Down è relativamente frequente e i pazienti che ne sono affetti hanno un maggior rischio di sviluppare leucemie acute, ma non tumori solidi.

Studi su grandi numeri hanno, infatti, evidenziato come i tumori solidi, tra cui i linfomi, siano, significativamente meno frequenti nei pazienti Down rispetto ai linfomi in bambini e adulti non affetti da trisomia del cromosoma 21. I linfomi testicolari, sono, comunque, rari ed estremamente aggressivi.

L'ecografia, associata a color Doppler, riveste un ruolo essenziale nella diagnosi e nella valutazione dei risultati delle terapie di queste lesioni.

Per la loro rarità, abbiamo ritenuto opportuno realizzare questo mini-pictorial essay.

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Introduction

Down's syndrome is relatively common, and in the United States one out of every 733 live births is affected. Patients with Down's syndrome have an increased risk of developing

acute leukemia, but not solid tumors [1]. According to various studies reported in the literature, some solid tumors [2] including lymphomas [3] are more frequent in patients with Down's syndrome, but studies performed on larger patient populations have shown that these tumors are significantly less frequent in Down patients than in children and adults who are not Trisomy 21-affected [1]. It is therefore possible that an extra copy of chromosome 21 has a protective effect with respect to the formation of solid tumors.

Testicular lymphomas are rare and account for 1%–2% of non-Hodgkin's lymphomas and 5% of testicular tumors with

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Figure 1 Testicular lymphoma in a 16-year-old patient with Down's syndrome. US shows hypoechoic intra-testicular lesions (arrows) (A); the deep lying lesion (arrows) (B) is hypervascular at color Doppler examination.

Figure 2 Testicular lymphoma in a 14-year-old patient with Down's syndrome. The testicle appears enlarged compared to the contralateral testicle; ultrasound shows numerous and voluminous hypoechoic areas (A) and hypervascularity at color Doppler examination (B, C).

an incidence of 0.26 per 100,000 people/year. They are extremely aggressive tumors with survival ranging from 12 to 24 months from the time of diagnosis [4]. They are generally cases of secondary localization of lymphoma and less frequently primary lymphomas.

The most typical clinical symptoms are pain and swelling, but these tumors may also, less commonly, cause fever, fatigue and loss of appetite.

Testicular neoplasm is diagnosed by ultrasound (US), whereas staging relies mainly on positron emission

tomography (PET) and computed tomography (CT) [4] while histological classification is performed on biopsy specimens; biopsy should be carried out under general anesthesia.

Unlike testicular masses, extra-testicular swellings are often benign [5]. The most common lesion is lipoma, while malignant lesions are much rarer including leiomyosarcoma, rhabdomyosarcoma, malignant fibrous histiocytoma, mesothelioma and lymphoma.

Clinical symptoms as well as US appearance of lymphoma of the tunica vaginalis testis are similar to those of testicular

Figure 3 Lymphoma of the tunica vaginalis testis in an 11-year-old patient with Down's syndrome. US shows significant thickening of the sheaths of the testis, indistinguishable from each other, with an inhomogeneous mainly hypoechoic echostructure (A). Color Doppler shows hypervascularity within the lesion (B,C).

Figure 4 Same case as Fig. 3 after one cycle of chemotherapy. Color Doppler shows reduced blood flow within the lesion which is a sign of positive response to therapy.

lymphoma, and both lesions are characterized by pain and swelling. Specificity of magnetic resonance (MR) imaging is elevated in lipomas but low in lymphomas and in the determination of the nature of the mass, which also in this case relies on biopsy.

US appearance

Among the various imaging modalities, US is the method of choice in the study of the scrotum. The literature reports values of sensitivity reaching 98%–100% [6,7] in the identification of scrotal masses. US examination is performed with the patient in the supine position; the scrotum is suspended by a support device and the scrotal sac is smoothed by gentle traction of the penis towards the navel, exerted by the patient using his left hand [8].

High frequency probes (9–18 MHz) are used, and orthogonal scans, usually axial and coronal, are obtained on the entire scrotum. The examination is completed using color Doppler evaluation.

Color Doppler parameters must be optimized for the assessment of slow flow (low pulse repetition frequency (PRF), wall filters set at minimum values, gain set to maximum in the presence of artifacts and the US beam focused on the region of interest) [8].

Under physiological conditions, color Doppler will show a rich vascularization of the testicle but no vascularization of the epididymis and tunica vaginalis testis. At B-mode US examination, the testicle presents a homogeneous US structure whereas the epididymis is slightly hyperechoic compared to the testicle, and the sheaths of the testis are visible.

At US, testicular lymphomas, single or multiple, unilateral or bilateral, appear as rounded hypoechoic formations (Figs. 1A and 2A), generally hypervascular (Figs. 1B and 2B,C) compared to normal parenchyma. The testicle may be enlarged [9].

When the sheaths of the testis are involved, the sheaths of the testis appear indistinguishable from each other, enlarged with an inhomogeneous, mainly hypoechoic echotexture (Fig. 3A). At Color Doppler examination the involved area appears hypervascular [10] (Fig. 3B, C).

US has an important role in the diagnosis of testicular neoplasm [11] and also for assessing treatment results. At follow-up after chemotherapy, in the presence of positive response to therapy, US will initially show reduced vascularization (Fig. 4) and only later a reduced volume and alterations in the echo pattern.

Discussion and conclusions

Lymphomas of the testicles and the tunica vaginalis testis are rare and account for 1%–7% of all tumors of the scrotal area, and they are even rarer in patients affected by Down's syndrome [4]. Like all other malignant lesions of the scrotum, lymphomas are characterized by pain and swelling. They are diagnosed by US and biopsy [9,11]. Staging generally involves clinical examination, laboratory tests, US examination and PET/CT, and the staging pattern is identical to that of lymphomas in other regions [12].

Treatment of choice in testicular lymphoma is orchiectomy, but the usefulness of chemotherapy after surgery is still a matter for debate, whereas treatment of lymphoma of the tunica vaginalis testis is basically chemotherapy [11]. In both types of cancer, prognosis is poor, and survival from time of diagnosis does generally not exceed 2 years [4].

It was decided to publish this mini-pictorial essay due to the rarity of the lesions and in order to highlight the role of ultrasound in the diagnosis and monitoring of treatment results.

Conflict of interest statement

The authors have no conflict of interest to declare.

Appendix

Supplementary data

Supplementary data associated with this article can be found, in the online version, at [doi:10.1016/j.jus.2011.07.001](https://doi.org/10.1016/j.jus.2011.07.001).

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