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Case Report

Cyst of canal of Nuck in a young woman affected by kniest syndrome: ultrasound and MRI features

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ABSTRACT

Swelling of the inguinal region in a woman is a frequent finding in daily clinical practice. We focused our attention on the possible differential diagnoses, giving emphasis to the less common causes of this frequent female disorder and describing their characteristics in ultrasound and magnetic resonance imaging. The causes of swelling in this region may be multiple and in this paper, we will show a rare case of diagnosis of Nuck's canal cyst in a patient with Kniest Syndrome.

Case report

A 37-year-old patient affected by Kniest syndrome (a rare polymalformative syndrome caused by the mutation of the COL2A1 gene on chromosome 12, characterized by dwarfism, visual disturbances, and auditory disorders) presented at our emergency department with a sore swelling in the right inguinal region.

From October to December 2017, the patient noticed rapid enlargement and increasing pain of that region. Upon interrogation, she denied urinary disturbances or intestinal irregularity.

The patient was referred to clinical examination, laboratory tests, and ultrasound evaluation.

Physical examination revealed a tense-elastic consistency lump of 3 cm in size, extending along the inguinal region, with smooth surface, soft consistency and slight fluctuation, not reducible, and modestly tender on palpation. The overlying skin showed no signs of inflammation. Laboratory blood tests were within the limits: C-reactive protein 0.2 mg/100 mL (reference [ref]: 0.00-0.50 mg/100 mL), hemoglobin = 12.7 g/dL (ref = 12-16

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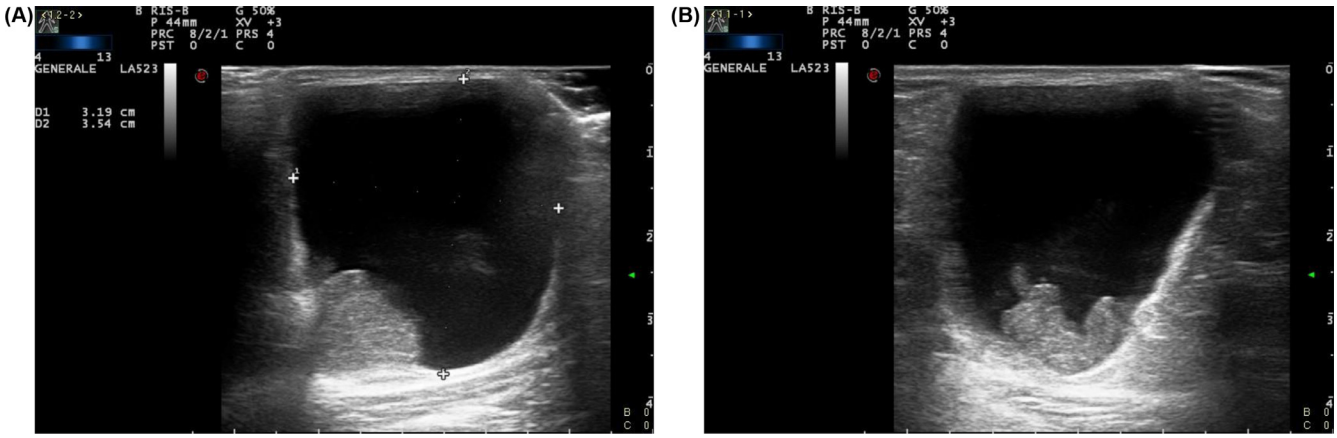

Fig. 1 – Ultrasound (US) evaluation (Philips Model No: iU22, Bothell WA) using a linear transducer (7.5-10 MHz) that revealed a tubular cystic structure of 3.1 x 3.5 cm in size with internal ipoecogenic mass. The evaluation with Color-Doppler module did not showed intralesional vascular signal. Lymphadenomegalies and other pathologic findings were not evident in the same region.

g/dL), and platelet count 237,000 u/L (ref: 156,000-373,000 u/L), velocity of erythrosedimentation 9 mm/h (ref: 0-21 mm/h).

We then performed an ultrasound evaluation (Philips Model No: iU22, Bothell WA) using a linear array transducer (7.5-10 MHz) that revealed a tubular cystic structure of 3.1 × 3.5 cm in size with an internal hypoechoic mass. Following Color-Doppler assessment did not show intralesional vascular signal.

The rest of the exam was unremarkable, with no evidence of local nodal enlargement or any other pathologic finding.

A magnetic resonance imaging (MRI) was subsequently performed to assess size, location, and morphostructural features of the lesion.

The MRI was performed with a 1.5-T MRI scanner (Intera 1.5 T; Philips, Best, The Netherlands) before and after intravenous administration of contrast medium (Gadovist 1.0 mmol/mL; Bayer, Munich, Germany) followed by saline chaser. We used T1-weighted, T2-weighted, and short tau inversion recovery sequences on axial, sagittal, and coronal planes prior to contrast administration, and 3D gradient echo T1 dynamic sequences.

The lobulated lesion presented low signal intensity in the T1-weighted sequences and high signal intensity in the T2-weighted sequences. Inner components had homogeneous signal intensity in all noncontrast sequences. Contrast-enhanced T1-weighted images showed no enhancement in arterial and venous phases.

The MRI features of the aforementioned mass, suggestive of a cystic lesion, lead to the suspected diagnosis of inguinal canal hydrocele. The patient was then subjected to excision of the mass and the histopathology diagnosis confirmed the hydrocele of the Nuck canal.

Discussion

A palpable mass in the female inguinal canal opens up to a variegated range of possible differential diagnoses that can only be defined thanks to the information provided by imaging and anatomopathologic data. Before discussing the most and least frequent causes of this condition, we must briefly review the embryological development of the female inguinal canal and the elements within.

The inguinal canal is a structure crossing the abdominal wall, lying on the medial half of the inguinal ligament, which in females conveys the round ligament of uterus along with the ilioinguinal nerve. It has a length of about 5 cm with an oblique course, directed anteroinferiorly, and medially. The inguinal canal has 2 openings: a superficial outlet called the subcutaneous inguinal ring (or superficial or external inguinal ring), and a deep one, called the abdominal inguinal ring (or deep or internal inguinal ring). The round ligament originates from the ovary gubernaculum, then enters the inguinal canal wrapped in a sheath of dependence of the transverse fascia of the abdomen, and reaches the external or subcutaneous orifice of the inguinal canal ending its course in the fat tissue of labia majora. In its path in the inguinal canal, the round ligament is accompanied by a fold of the parietal peritoneum, which actually constitutes the canal of Nuck.

During the development of the female fetus, obliteration of this peritoneal fold takes place; if only partial proximal obliteration occurs, with patency maintenance of the distal portion, a cyst of the Nuck canal is determined [1]. Otherwise, if it remains totally patent an indirect inguinal hernia is generated [2].

Palpable masses of the inguinal region may give rise to several possible differential diagnoses; in particular, the most frequent cause is an inguinal hernia in which omentum and bowel loops are displaced in the inguinal canal. Uncommon pathologic causes should also be considered such as nodal enlargements, soft tissue neoplasms, cysts of the ganglia, and perineal abscesses [1].

The imaging characteristics of the various pathologic conditions that may affect the inguinal region can guide the diagnostic suspicion toward the specific entity in question.

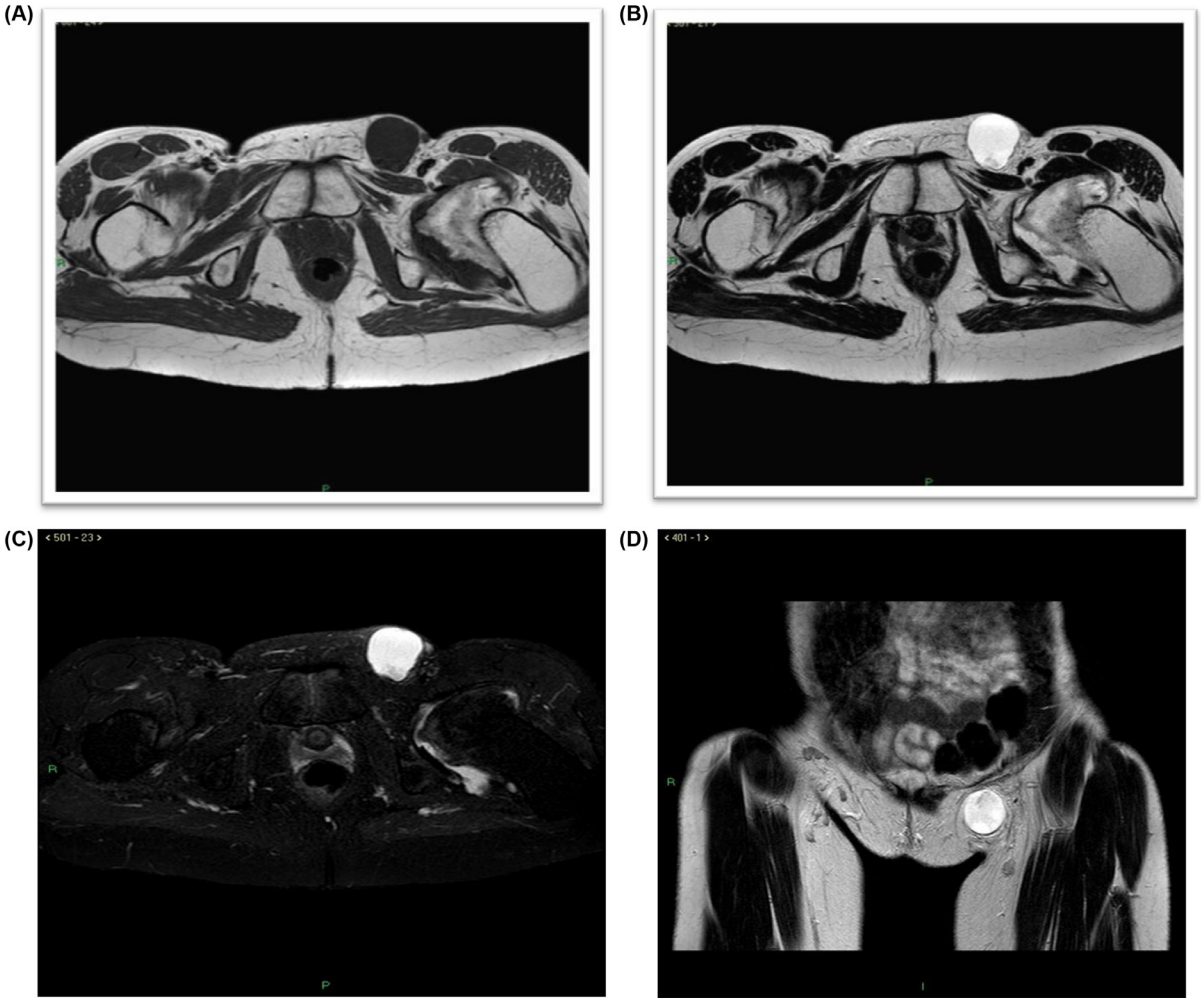

Fig. 2 – Axial, coronal MRI of the pelvis. Technique: 1.5-T MRI scanner, Sense Torso Coil. (A) Turbo spin echo (TSE) T1 sequence (axial plane). (B) TSE T2 sequence (axial plane). (C) TSE T2 sequence (coronal plane). (D) TSE STIR (axial plane). The lesion shows a low signal intensity in the T1-weighted sequences and high signal intensity in the T2-weighted sequences. Inner components had a homogeneous signal intensity in all noncontrast enhanced sequences. MRI, magnetic resonance imaging; STIR, short tau inversion recovery.

For example, an abscess of the inguinal region can be described as an irregular hypoanechoic area, with posterior acoustic enhancement and internal echogenic debris. The evaluation with Color-Doppler module documents peripheral hyperemia and absence of vascular signal inside [3]. MRI shows a hypointense in T1 sequences and hyperintense in T2 and short tau inversion recovery sequences lesion, as well as a pseudocapsule with low signal intensity in all sequences with "ring" enhancement after administration of intravenous paramagnetic contrast medium [3].

A ganglionic cyst appears as a multilocular, thick-walled, well-defined anechoic formation with hypervascular septa [4]. A lipoma, on the other hand, presents itself as a formation with regular margins and well-defined borders, sometimes outlined by a thin capsule. The echostructure may be iso-hypo or hyperechoic with the absence of vascular signals at Color-Doppler assessment [5]. Lipomas appear hyperintense in MRI images obtained with T1-weighted sequences with proportional reduction of the signal in T2-weighted images with fat signal saturation.

A liposarcoma is a mass with irregular margins, possible lobulations, and a nonhomogeneous ecostructure with peri and/or intralesional vascularization. On MRI, liposarcoma appears as a lesion with an intermediate-low signal in the T1 sequences, high signal in the T2 sequences, and enhancement after gadolinium administration. In fast-growing lesions, a central necrotic core also may be present [6].

However, a lump in the inguinal region could also be a lymph node, showing a well-recognizable hilar structure and typical morphology, possible expression of a loco-regional inflammatory process. Sometimes we may find multiple lymph nodes with globose morphology and a central necrotic area. They are indicator of malignant processes, lymphoproliferative syndromes, or sometimes infectious diseases.

The sonographic finding of an anechoic pulsating inguinal mass, in communication with the femoral artery, must direct the diagnostic suspicion toward a pseudoaneurismatic formation. In this case, ultrasound with Doppler is crucial in the diagnosis, showing prominent blood flow, with an arterial pattern and a reversed flow component usually described as "to-and-fro" [7].

Eventually, a canal of Nuck cyst can be taken into account as a possible diagnosis of inguinal region swelling.

In literature, only 400 cases of hydrocele of the Nuck canal are described [8]. In one-third of the patients, this finding may be associated with inguinal hernia, therefore, some authors have proposed the Valsalva maneuver during the ultrasound examination to exclude or confirm its coexistence [9]. Sonographically, the Nuck canal cyst presents as an anechoic tubular or oval lesion in the inguinal region or in the labium major [10].

We finally considered that our patient was affected by Kniest Syndrome, a rare disorder that arises from a genetic mutation of the COL2A1 gene. Clinically, it is characterized by short-trunked dwarfism, kyphoscoliosis, and enlarged joints with restricted mobility. Other features include marked hand arthropathy, cleft palate, hearing loss, and ocular abnormalities (myopia, abnormal vitreous, and high risk of developing retinal detachment) [11,12].

To the best of our knowledge, there are no data in literature that reported clinical association between this polymalformative dysplasia and the Nuck canal cyst.

Conclusion

A Cyst of canal of Nuck canal must always be included in the differential diagnosis of cystic inguinal lumps in women. Clinical data, medical history, ultrasound, and MRI imaging are useful tools in obtaining a correct diagnosis, albeit surgery is needed for confirmation (Figs. 1-2).

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