Case report: Inflammatory myofibroblastic tumor of pancreatic origin in a patient with down syndrome: The role of diagnostic ultrasound

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KEYWORDS

Inflammatory myofibroblastic tumor (IMT);
Nonsteroidal anti-inflammatory drugs (NSAIDs);
Down's syndrome;
Computed tomography (CT).

Abstract *Introduction:* Inflammatory myofibroblastic tumor (IMT) is a benign solid tumor of uncertain etiology.

Materials and methods: We report a case of a 4-year-old Down syndrome affected child, who had a pancreatic mass identified by ultrasonography (US) and confirmed by computed tomography (CT).

Results: Monitoring of IMT was performed by serial US studies, and at follow-up after 4 years there was no relapse.

Discussion: As radical removal of the lesion was not possible, the patient was successfully treated with nonsteroidal anti-inflammatory drugs (NSAIDs). It was decided to monitor the lesion by serial US in order to reduce the number of CT examinations and thereby avoid excessive exposure to ionizing radiation. It is widely reported in the literature that repeated CT scans are associated with increased exposure to radiation which may cause cancer, a fact which should not be overlooked in children.

Sommario *Introduzione:* Il tumore miofibroblastico infiammatorio (TIM) è un tumore solido benigno, di incerta eziologia.

Materiali e metodi: Riportiamo un case-report di un bambino (4 anni, sindrome di Down), con massa pancreatica identificata all'ultrasonografia (US) e confermata da TC.

Risultati: Il monitoraggio del TIM è stato effettuato con US seriate. Il follow-up a 4 anni dalla diagnosi è senza recidive.

Discussione: Il paziente, data l'impossibilità dell'exeresi chirurgica radicale, è stato trattato con successo con terapia medica (FANS). Abbiamo deciso di monitorizzare la lesione con ultrasonografie seriate, riducendo il numero di esami TC ai quali il paziente avrebbe dovuto sottoporsi, evitando così l'esposizione a radiazioni ionizzanti. La letteratura, infatti, riporta ampiamente come l'esecuzione di ripetuti esami TC si accompagni ad una importante radioesposizione con conseguente rischio cancerogeno, dato assolutamente non trascurabile in età pediatrica.

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Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare solid lesion, most frequently located in the lungs and rarely in the pancreas [1]. Pancreatic masses account for 0.2% [2] of all pediatric cancers and IMT can be clinically and radiologically indistinguishable from malignant lesions. The most frequently proposed etiological hypotheses include immunological, infectious or traumatic causes [3]. Surgical resection is the therapeutic gold-standard, but we present a case of IMT of pancreatic origin which was successfully treated with NSAIDs [4]. After this therapeutic approach, instrumental monitoring and follow-up was carried out exclusively by US.

Case report

A 4-year-old Down syndrome affected boy was hospitalized for abdominal pain without vomiting. Clinical examination was negative. Abdominal US (Fig. 1) showed a hypo-hyperechoic inhomogeneous mass (4×3 cm) involving the head of the pancreas. The mass was adjacent to the inferior vena cava, and satellite lymph nodes were increased in volume (1.5–2 cm). Computed tomography (CT) (Fig. 2) confirmed a hyperdense round solid lesion involving the head of the

Fig. 1 US showing a mass in the pancreatic head.

Fig. 2 CT scan showing a mass in the pancreatic head.

pancreas and associated with dilatation of Wirsung's duct in the distal tract and moderate dilatation of the intrahepatic bile ducts and the common bile duct. Laboratory test results were abnormal: erythrocyte sedimentation rate (ESR) 53; C-reactive protein (CRP) 37.50; glutamyl oxaloacetic transaminase (GOT) 215; glutamate pyruvate transaminase (GPT) 475; blood bilirubin 3.56 mg/dl and urinary bilirubin 1.0 mg/dl; tumor markers were negative. The patient underwent surgical biopsy, and histological examination showed a morphological pattern compatible with IMT of pancreatic origin. Surgical removal of the lesion was not possible without damaging the surrounding tissues, and medical treatment with NSAIDs was therefore preferred, as reported in the literature [4]. In the following days the symptoms cleared up: the patient passed normocolic stool, urine was normochromic and laboratory test results returned to normal. CT scan (Fig. 3) two months after diagnosis showed that IMT was reduced by one half. The patient underwent monthly US examinations (Fig. 4) for the duration of the medical therapy. He subsequently underwent monthly clinical examinations and laboratory tests in addition to abdominal US examination every six months. US showed volume reduction of the mass until complete disappearance. Follow-up 4 years after diagnosis showed that the patient was free of disease.

Fig. 3 CT scan at the 2-month follow-up showing a reduced mass in the pancreatic head.

Fig. 4 US in the early follow-up showing a reduced mass in the pancreatic head.

Written informed consent was obtained from the patient's parents for the publication of this case report and accompanying images.

Discussion

IMT is an atypical tumor which may not be easy to distinguish from a malignant tumor due to its invasiveness and tendency to recur [1], and US, CT and magnetic resonance imaging (MRI) can often not provide a definitive diagnosis [6]. Radical surgery is the gold-standard [1], but surgical treatment is not always possible. Among the proposed alternative medical treatment approaches [5], the recent literature contains reports concerning the use of NSAIDs [4]. In the present case, radical resection was not possible, and the patient therefore received ibuprofen (200 mg/day). The size of the tumor gradually decreased and it eventually disappeared. Given the close correlation between the US and CT images, and particularly the non-invasiveness of the US technique compared to CT not only in terms of radiation

exposure, but also the fact that sedation is not required whereas it is obligatory in pediatric patients who undergo CT, the patient was treated conservatively, from a therapeutic point of view, and was therefore monitored by US alone. Medical therapy was administered for 6 months. The 4-year follow-up showed that the patient had no recurrence and was free of disease.

Conflict of interest statement

The authors have no conflict of interest.

Supplementary material

Supplementary data associated with this article can be found in the online version at [doi:10.1016/j.jus.2010.11.003](https://doi.org/10.1016/j.jus.2010.11.003).

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