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Pitfalls in the follow up for appendiceal carcinoid in a girl: a case report.

Marco Gasparella[1], Patrizia Dall'Igna[1], Gianni Bisogno[2], Pietro Zucchetta[3], Isolde Unterkalmsteiner[4], Paolo Andriolo[4], Rita Alaggio[5], Giovanni Cecchetto[1]

[1]Pediatric Surgery Clinic, Pediatrics Dipartment, University of Padova

[2]Division of Pediatric Ematology-Oncology, Pediatrics Dipartment, University of Padova

[3]Department of Nuclear Medicine I, University-Hospital of Padova

[4]Division of Pediatric Surgery, Hospital of Bolzano

[5]Oncologic Sciences Institute, Dipartment of Pathology, University of Padova

Address for Correspondence:

Marco Gasparella

[1]Pediatric Surgery Clinic, Pediatrics Dipartment, University of Padova

Via Giustiniani 1, 35100 Padova

Telephon number: 049-821-3681; Fax: 049-821-1781.

e-mail: marco.gasparella@unipd.it

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Introduction

Carcinoid tumors of the gastro-intestinal tract are rare tumors, usually incidentally discovered after an operation for different reasons. Those located in the appendix are the most common gastro-intestinal epithelial tumors in childhood (1), and are always found after an operation for acute appendicits (2).

The behaviour of these neoplasms in children is less aggressive comparing it with that of adults: the lesions are generally small, not invasive and not characterized by distant metastases (3). Due to their rarity and the evidence in literature of only a few complicated cases, the clinical-therapeutic approach still represents a challenge. In this report it is discussed a case of a 15 year old girl, who presented hepatic nodules simulating metastatic lesions during her follow-up for appendiceal carcinoid (AC).

Case report

A carcinoid of the appendix was discovered in a 15 year old girl, after appendectomy. The tumor, which measured 3 mm, was localized in the tip and did not showed invasion of the serosa and periappendix fat.

As recommended by the treatment guidelines of the Italian Study on Rare Tumors (4), after the operation, she entered a yearly follow-up with the urine dosage of 5-hydroxyndolacetic acid (5-HIAA) and abdominal ultrasonography (US). Three years after, while the 5-HIAA was negative, the US demonstrated a 15 mm solid nodule in the first segment of the liver, and the girl underwent a further evaluation: a CT scan confirmed the presence of the nodule (figure 1), and then, according to the more recent guidelines, it was decide to evaluate the seric dosage of Chromogranine A, which was negative, and to perform an Octreotide scintiscan to rule out AC metastases .

Beside the nodule in the segment I, this investigation demonstrated other two positive lesions in the V and VIII hepatic segments (figure 2). These findings were in contrast with the total absence of symptoms and signs related to a carcinoid syndrome, however the positivity of the Octreotide scintiscan was suggesting possible metastases. Therefore a surgical exploration was decided: through a laparoscopic approach, only the nodule in the I segment could be visualized and the procedure was converted: however, even touching the liver, and under the intraoperative ultrasound guidance, the other two nodules could not be identified. The procedure was concluded with the excision of the visible lesion and the biopsy of some lymph-nodes. The histological diagnosis was in favour of focal nodular hyperplasia and the lymph-nodes were normal. The patient continued her normal follow-up, and repeated an Octreotide scintiscan two years after the laparotomy, which confirmed the nodules in the V and VIII segments, but with a less intensive captation At present , after 6 years of follow up, the girl remains asymptomatic.

Discussion

An appendiceal carcinoid is generally diagnosed in the fourth-fifth decade of life. The precise incidence in children is not known but a summary of all publication yelds a frequency of 2-5 per 1000 appendectomies (3).The best therapeutic behaviour after detection of a AC in children is still debated. While appendectomy alone has been considered for tumors < 2 cm, the need of a right colectomy for tumors > 2 cm, traditionally accepted (5), remains nowadays controversial, and recent experiences favour a non-aggressive approach (6). Moreover, the invasion of serosa and periappendiceal fat is not considered as an unfavourable factor.

In all cases a clinical work-up to detect regional or distant metastases is recommended after the appendectomy. Considering the limited experience on the clinical behaviour of this tumor in childhood, the investigations adopted for pediatric patients are similar to that suggested for adults. Beside the urine dosage of 5-HIAA and US, the scintiscan with Octreotide, a somatostatine analogue, has been introduced in the general work-up, since AC express somatostatine receptors (7). Nowadays this is considered the most important diagnostic procedure.

In adults, the liver represents the most frequent site of metastases, but in childhood distant metastases, and consequent symptoms and signs of a carcinoid syndrome, have not been observed so far (3).

In our patient, the tumor was small and not invasive at histology, and the clinical-radiological evaluation after appendectomy was negative for other localizations; in spite of these favourable features, the lesion found in the liver after 3 years was strongly suggestive of a liver metastasis of AC, even if the urine dosage of 5-HIAA had remained negative. The positivity of the Octreotide scintiscan made us to consider a surgical exploration.

The histological diagnosis of FNH was a surprise in our case, but it is not an unusual finding in childhood and adolescents: 15% of FNH occurs in patients whose age is comprised between 1 and 16 years, and the incidence in females, comparing it with that of males is 5:1 (8) In adolescents, FNH is usually incidentally observed at US performed for other reasons, however it may manifest as an hepatic mass. In most of the cases, the history of the patient (i.e. oestroprogesterone assumption) and the radiological characteristics may address the diagnosis. Histological evaluation is mandatory to confirm the nature of the lesion, for the differential diagnosis with a malignant tumor. The positivity of the Octreotide scan in case of FNH is possible because of the presence of activated lymphocytes as happened in our case (figure 3), but this exam is not utilized for the diagnosis of FNH, because unspecific for this lesion (9).

The histological response and the further follow-up in our patient confirm the good prognosis of AC of the appendix, especially those of small size.

The litterature data, based overall on adult series, stress a long follow-up, because local recurrences have been observed many years after the appendectomy (10). However discussion may arise in cases of small tumors, that usually have a benign behaviour, especially in pediatric age: in our patient the diagnostic and therapeutical approach, considering the size of the tumor, might have been too aggressive.

Conclusion

The question is whether or not the follow-up in children who have small, non invasive tumors, should be limited just to the dosage of 5-HIAA, avoiding stress and unuseful procedures on one hand, and costs on the other. It could be interesting to understand, moreover, if the Octreotide scintiscan, that is highly specific in adults, has the same significance in children, considering that lesions containing lymphocytes may result hyper-uptaking. Multicentric studies on larger series could obtain more clinical data and establish in the future a more adequate approach.

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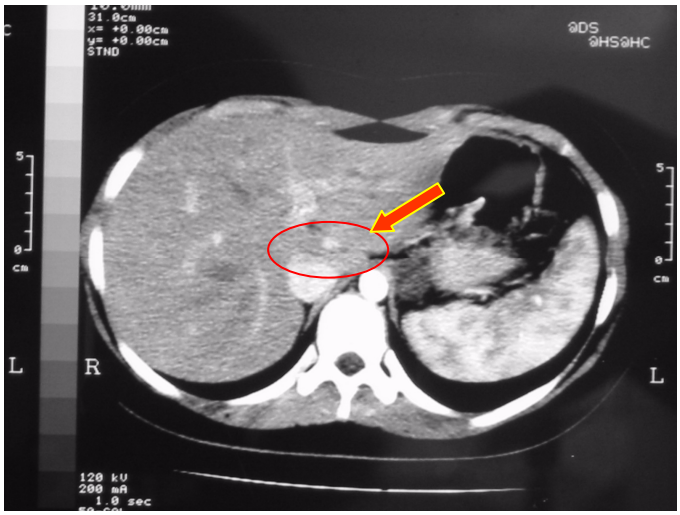
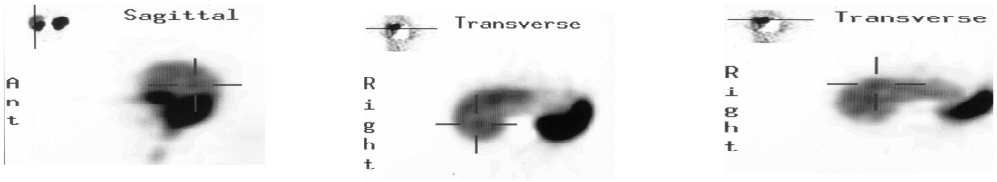


Figure 1: CT scan confirmes the presence of the nodule in the first segment of the liver (red arrow).



A) **B)** **C)**

Figure 2: Octreotide scintiscan: the positivity of three lesions in the I (**A**), V (**B**) and VIII (**C**) hepatic segments.

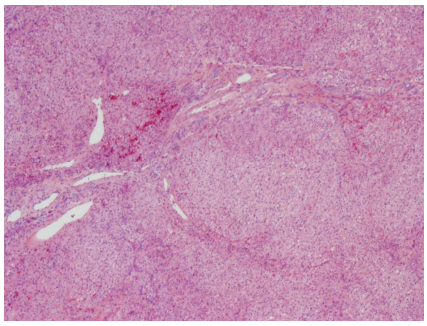


Figure 3: He Stain: Central stellate fibrosis contrining a large vessel and numerous lymphocytes (Ematossilina-Eosina, 40X).