STROMAL TUMOR OF THE AMPULLA OF VATER: REPORT OF A CASE AND SYSTEMATIC REVIEW OF REPORTED CASES.

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LES TUMEURS STROMALES DE L'AMPOULE DE VATER : REVUE SYSTÉMATIQUE DES FAITS CLINIQUES RAPPORTÉS DANS LA LITTÉRATURE.

LA TUNISIE MEDICALE - 2009 ; Vol 87 (n°09) : 556 - 559

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RÉSUMÉ

Prérequis : Les tumeurs stromales digestives regroupent plusieurs lésions néoplasiques non épithéliales se développant aux dépens de la paroi du tractus digestif. La localisation au niveau de l'ampoule de Vater est très rare.

Le but de cette étude est de décrire les particularités clinique, endoscopique et thérapeutique des tumeurs stromales de l'ampoule de Vater.

Methodes : Les auteurs rapportent un nouveau cas et réalisent une revue de la littérature en utilisant Medline. Les mots-clés utilisés sont ''ampoule de Vater'', "tumeur stromale digestive" et "CD 117". En plus de ce nouveau cas, sept autre cas ont été retrouvés dans la littérature.

Resultats :L'analyse de ces huit cas permet de tirer les conclusions Les tumeurs stromales de l'ampoule de Vater touchent les adultes de la cinquantaine. Il n'y a pas de symptomatologie spécifique. La fibroscopie digestive haute avec biopsie et immunohistochimie a permis de poser en pré opératoire le diagnostic positif dans 5 cas. Le traitement devrait être une duodénopancréatectomie céphalique car la tumeur est souvent maligne (5cas).

MOTS-CLÉS

Ampoule de Vater, Tumeur stromale digestive, CD 117.

SUMMARY

Background : Gastrointestinal stromal tumours are a large category of primary no epithelial neoplasms of the digestive tract. The localization of stromal tumours in the ampulla of Vater (STAV) is very rare.

Aim : The aim of this study was to describe clinical, endoscopic and therapeutic particularities of STAV.

Methods : We reported a new case and we carried out an extensive electronic search for the relevant literature using Medline. Key words used were "ampulla of Vater" and "Gastrointestinal stromal tumor" and "CD 117". With our case, we collected seven other cases in the literature.

Results : The analysis of the sample of these eight cases leads us to propose the following conclusions. STAV is a tumor of adult after the age of fifty. There is no specific symptomatology. Gastroduodenal endoscopy with biopsies and immunoassaying allows positive preoperative diagnosis in the 5 cases. Treatment should be duodenopancreatectomy since the tumor is often malignant (5 cases).

KEY-WORDS

Ampulla of Vater - Gastrointestinal stromal tumor - CD 117.

Gastrointestinal stromal tumours (GISTs) are a primary no epithelial neoplasms of the digestive tract. Its prevalence ranged between 0.1 to 3 % of gastrointestinal tumours [1]. They are localised in the stomach (60 %) and the small intestine (20 to 30%) [2]. They probably derive from the Cajal cell which is essentially located in the sub mucosa and express CD 117 also called c-kit [3]. The localization of GIST in the ampulla of Vater is very rare. The aim of this study was to report a new case of stromal tumor of the ampulla of Vater (STAV) combined with a systematic review of reported cases published in peer-reviewed journals.

MATERIAL AND METHODS

A new case of STAV was reported. An extensive electronic search for the relevant literature was carried out using Medline. Key words used were “ampulla of Vater” and “Gastrointestinal stromal tumor” and “CD 117”. Only articles reporting one or several cases were taken into consideration. Reviews of the literature, systematic reviews, letters to editors, and abstracts were excluded. A descriptive analysis of the collected samples including our case was performed. Morphological, therapeutic and outcome variables were then reported.

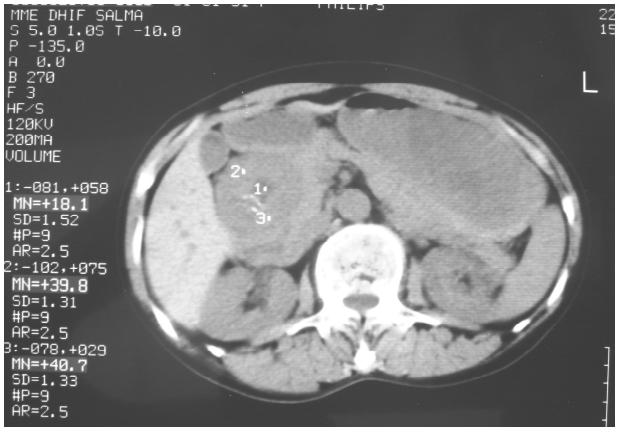
RESULTS

Our case

A 55 year-old woman presented with a three-year history of epigastric pain, weakness and weight loss. Her past medical history was unremarkamable. The physical exam showed a fixed epigastric mass measuring 5 cm without pain. Lab tests were normal.

Ultra sonography (US) showed a 5 cm hypoechogenic image that had lobulated limits and contained central calcifications. It was pushing back the duodenum, the inferior vena cava and the mesenteric vessels. It was impossible to determine whether it was a pancreatic or hepatic mass. Computed tomography (CT) showed a solid, hypodense pancreatic mass with a peripheric enhancement that contained central calcifications (Figure 1). Barium swallow revealed an enlargement of the duodenum (Figure 2).

Figure 1 : Computed tomography: Solid and hypodense mass containing central calcifications that enhances at the periphery



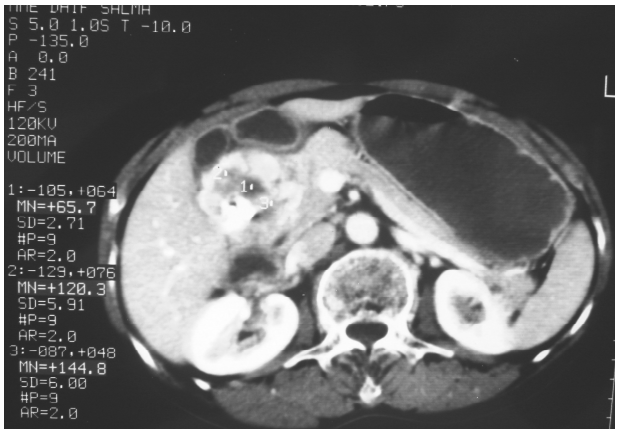
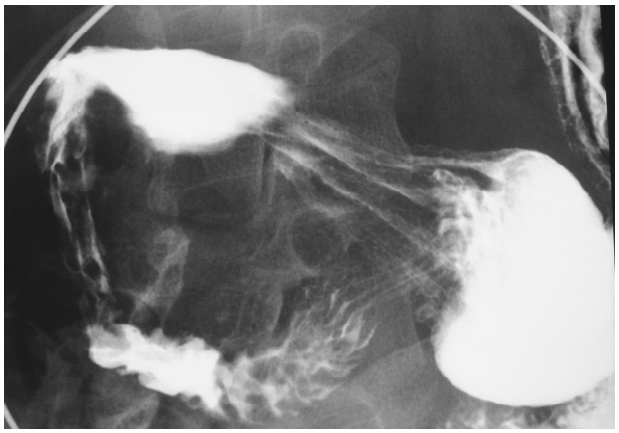


Figure 2 : Barium swallow: Enlargement of the duodenum



Based on these preoperative findings, the diagnosis was an exocrine tumor of the head of the pancreas. There was no evidence of either extra pancreatic or peritoneal metastasis.

The patient was operated on July, 30th 2006. Intra-operative exploration found a cephalic pancreatic tumor. A Whipple procedure was performed.

Grossly, the tumor was white and nodular with areas of necrosis and measured 4 x 4.5. It arises from the ampulla of Vater. The pancreas was not involved by the tumor. Microscopic and immunohistochemical studies revealed spindel-shaped cells that were positive for CD 117 and Vimentin. Mitoses were rare. The final diagnosis was benign STAV.

The post-operative course was uneventful and the patient left the hospital 10 days after operation.

One year later, she was asymptomatic. Ultrasound and CT performed six months after were normal.

Descriptive analysis of the seven cases reported with our case (Table 1):

Table 1 : Description of cases collected in the English medical literature and our case

reference	authors	sex	age	sign/symptoms	physical examination	endoscopy	biopsy	ERCP	EUS	CT scan	intervention	size of tumor	malignancy
4	Koçer N.E	M	44	weight loss	-	+	GIST	+	no	+	wheeple	9	yes
5	Singhal D.M	M	69	melena	pallor	+	negative	no	+	+	wheeple	3	no
6	Filippou D	F	65	weakness jaundice	jaundice	+	GIST	+	no	+	local resection	6	yes
7	Takahashi Y	F	77	-	pallor	+	GIST	+ (bleeding)	no	+	wheeple	4	yes
8	Kim S.H	F	37	weakness melena /nausea	pallor	+	GIST	+	no	+	wheeple	5.5	yes
9	Moss AC	M	57	melena	-	+	GIST	no	+	+	local resection	2.6	no
10	Matsushita M	M	44	jaundice	jaundice	+	mesenchymal tumor	no	no	+	no (died)	8	yes
our case	Jerraya H	F	55	epigastric pain weakness/weight loss	epigastric mass	no	no	no	no	+	wheeple	4.5	no

Seven cases were collected from peer-reviewed journals. The diagnosis of stromal tumor of the ampulla of Vater was retained after pathologic examination of resected specimen in seven cases [4-9] and in one case, the diagnosis was based on postmortem examination [10].

There were four men and four women aged between 37 and 77 years with a median age of 56. Six patients presented digestive signs: melena in three cases [5,8,9], jaundice in two cases [6,10], abdominal pain in one case and nausea in one case [8]. Three patients complained of weakness and fatigue and two of weight loss. In one case, STAV was revealed by anemia [7].

Gastroduodenal endoscopy was performed seven times [4-10]. In six cases, it showed an ulcerating tumor of the duodenal ampullary region. In the remaining case, it showed bulging ampulla of Vater that bled on touch. The study of biopsies collected endoscopically from the ulcerating lesions suggested the diagnosis of GIST five times [4,6-9] and that of a malignant mesenchymal tumor once [10]. The biopsy specimen of the non-ulcerating lesion was only suggestive of inflammatory cells [5]. ERCP was attempted four times [4,6-8]. In two cases, it showed an ulcerated mass in ampulla. In the other cases, it was unhelpful and complicated by bleeding in one patient [7]. EUS was performed twice. It was highly suggestive of GIST in one case [5] by showing a hypoechoic tumor which originated from the muscularis propria. In the other case [9], it showed a hypoechoic and slightly heterogeneous mass which may be consistent with GIST.

US was performed twice. It only showed a dilatation of intra and extra hepatic biliary tract and pancreatic duct, without identifying the tumor. In the other case, it revealed a hypoechogenic lesion with central calcifications. Computed tomography was performed in all cases. It allowed individualizing the stromal tumor as a solid mass that enhanced uniformly or at the periphery in six cases. CT located the tumor as developing in the ampulla of Vater in two cases. Barium swallow, performed twice, was useless. Celiac and supramesenteric angiography, performed in only one patient, showed a hypervascular nodule.

All these diagnostic tools helped to evoke a stromal tumor of the ampulla of Vater in six cases. The positive diagnosis of mesenchymal tumor of the ampulla of Vater was suggested in one case [10].

Five patients underwent a Whipple procedure. Two patients were treated by local resection of the tumor via duodenotomy; one did not accept pancreatodudenectomy [6] and the second had a small tumor with no evidence of histological invasion [9]. The last patient died before operation [10].

The tumor size was superior to 5 cm in four cases. The tumor was benign in three cases. In the other cases, it was malignant: histological signs of malignancy were found five times but metastases were present only once.

DISCUSSION

The analysis of a sample of 8 cases (7 cases collected in the medical literature and our case) allowed us to conclude that STAV is a tumor of adult after the age of fifty. There is no specific symptomatology. Gastroduodenal endoscopy with biopsies and immunoassaying allows positive preoperative diagnosis in the majority of cases. Treatment must be duodenopancreatectomy since the tumor is often malignant.

STAV is a tumor of adults of the fifth decade which is in concordance with others histological types of ampullary tumours [11]. But it seems to be four times more frequent in women than it is in men while adenocarcinoma of the ampulla of Vater are significantly more frequent in men with a sex ratio of 1.5 [11]. The tumor is usually big at time of diagnosis with a size of more than five cm in four cases.

STAV presents a large clinical spectrum mimicking symptoms of adenoma and adenocarcinoma of the ampulla of Vater. It can be responsible for epigastric pain, jaundice with

hyperbilirubinemia, gastrointestinal bleeding with anaemia, nausea and vomiting. Weakness, fatigue and weight loss are frequent.

Radiological investigations usually performed to explore a pancreatic mass and jaundice have a diagnostic low-yield. In fact, in this review, US was unable to show the tumor in any case. Barium swallow, ERCP and celiac and supramesenteric angiography were useless. However, CT, which had a poor diagnosis role in adenocarcinoma of the ampulla as it showed the tumor in only 25% of cases [12], seemed to be helpful: it revealed STAV in the majority of cases (6/8) and was able to identify the tumor as an ampullary one in one third of them.

Positive preoperative diagnosis was carried out by gastroduodenal endoscopy with biopsy of the tumor. An ulcerating tumor of the duodenal ampullary region must evoke STAV. Histological exam of endoscopic specimen with immunoassaying confirmed the diagnosis or at least pointed a mesenchymal tumor. In our review, each time biopsies were performed (seven times), they were beneficial to preoperative diagnosis by showing a GIST in five cases and mesenchymal tumor in one case. Only once the histological exam was useless. EUS seems to be helpful as it showed the tumor, localized it at an ampullary one and evoked a GIST in the two cases it was performed .It will be for certain very helpful to improve result of biopsy.

Like other ampullary tumours, STAV are often malignant according to Douglas and al criteria of malignancy [13]. In our study, this is due to the high tumor size that was superior to five cm in four of the reviewed eight cases. All of them were malignant.

In 7 cases, a Whipple procedure was retained as treatment by the medical team. It was performed five times with an uneventful postoperative course in all cases. A wide local resection of the tumor via duodenectomy was performed twice. One patient refused duodenopancreatectomy, the other had a little tumor of 2.5 cm.

CONCLUSION

STAV is a rare neoplasm which it should be evoked in the presence of an ulcerating tumor of the ampulla of Vater. The positive diagnosis is usually made by biopsy despite the localisation of the tumor in the sub mucosa. Since the high risk of malignancy, the treatment should be duodenopancreatectomy which can ensure resection with negative margin.

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