Case report

Sister Marie Joseph nodule and Krükenberg’s tumor: rare association revealing a gastric adenocarcinoma

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A B S T R A C T

It was about a 61-year-old patient who presented an indurated and painless umbilical swelling surrounded by ulcerated plaque and an abdominal arch progressing for 8 months. Abdomino-pelvic ultrasound and abdominal-pelvic CT scan revealed in right ovarian the presence of a mass with a cystic’s tissue, a thickness with a subcutaneous infiltration of the umbilical region and the diffuse infiltration of gastric parietal mesenteric fat. Histology of the ovarian mass confirmed Krükenberg’s tumor and that of the Sister Marie-Joseph’s nodule confirmed umbilical skin metastasis. The primary was a well differentiated gastric adenocarcinoma. The death occurred after the first chemotherapy treatment. The association of Krükenberg’s tumor with Sister Marie-Joseph’s nodule, rare and unusual, has been revealing of a gastric adenocarcinoma which up to now was unknown and asymptomatic.

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I n t r o d u c t i o n

Sister Marie Joseph’s (JMS) nodule is a cutaneous umbilical metastasis of gastro-intestinal tract cancer or gynecologic area \cite{1}. It represents a discreet and rare clinical sign, which does not show the existence only of a malignant visceral tumor, but which also reveals the bad prognosis of these cancers due to the late diagnosis \cite{2}.

Krükenberg’s tumor (KT) is a rare ovarian metastasis, more often bilateral and secondary to digestive cancer, predominantly gastric \cite{3}. It is characterized by muco-secreting \textless signet-ring\textgreater in ovary in 90% of cases \cite{3}. It represents only 1%-2% of ovarian tumors and often happen to genitally active women \cite{4}. It is often of fortuitous discovered during a digestive neoplasm assessment \cite{5}.

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Primary cancers, especially those who arise from stomach, are indeed small while ovary tumor is already big [6]. The association of Krükenber tumor to sister Marie Joseph’s nodule is rare. We report this metastatic association and unusual which revealed a gastric adenocarcinoma to a 61 years old patient without a particular medical history.

**Observation**

Sixty-one-year-old woman with no significant medical history was hospitalized in our internal medicine department for abdominal distension with general condition deteriorated for 8 months. Clinical examination found a palpable nontender nodule in umbilical area, surrounded by an ulcerated plaque (Fig. 1), and an abdominal swelling.

Abdominal ultrasonography showed a heterogenic cystic-tissue mass of the right ovary with some hyperechoic mural nodules (Fig. 2).

Abdominopelvic CT scan found an umbilical nodule thick with a sub-cutaneous infiltration of the umbilical area (Fig. 3). It also showed a thickening with a diffuse infiltration of gastric parietal mesenteric fat adjacent (Fig. 4) and a multi-compartmentalized cystic tissue mass on the uterine side with mural nodules.

The CA marker was at 347, 21 U/mL and the CEA was at 9, 51ng/mL. facing several radiologic signs, the problem of medical treatment was aroused. Multidisciplinary medical Staff decided to realize a biopsy. The ovarian masses biopsy found big signet-ring cells meaning of an ovarian metastasis of adenocarcinoma who’s the primary tumor had to be found.

Umbilical area biopsy found a metastasis of a gastric adenocarcinoma.

An upper endoscopy found a gastric nodule who’s histologic exam confirmed an adenocarcinoma well-differentiated.

Eventually our patient was suffering a gastric adenocarcinoma with cutaneous umbilical and ovarian metastasis.

Six chemotherapies were held but the death happened after 1 month of treatment.

**Discussion**

Metastases are secondary hotbed of cancer, developed far from the primary and who’s the growth is autonomous, independent from the primary cancer [7]. The mechanism of occurrence of these metastases in the natural history of a cancer is variable [8]. It is revealing of an asymptomatic primary tumor and therefore unknown. It can also be contemporary to the primary tumor discovered either during the extension assessment or because it causes clinical symptoms. In our study, these metastases were revealing of the primary tumor.

The JMS nodule is a skin metastasis from abdominopelvic cancer [8]. Its incidence is low, representing 1%-3% of all abdominopelvic neoplasms with a poor prognosis due to its late diagnostic, especially in a country with limited resources [1]. The gastrointestinal sphere is the most frequent site of the primary tumor in 35%-65% of cases followed by a gynecologic origin in 12%-35% [9]. The clinical presentation of the JMS nodule is often that of a general firm, indurated swelling with irregular margins. The surface is sometimes fissured or ulcer-necrotic; often painful or even itchy. The size is variable, from 0.5 to 2 cm in general [10]. Some few spectacular cases up to 10 cm have been reported [10]. In our case, the nodule presented itself as an indurated umbilical swelling surrounded by an ulcerated plaque. In 30% of cases, the JMS nodule is a discrete, and rare clinical sign representing one of the initial presentation patterns of visceral neoplasia [11].

The imagery is no specific [12]. It does not allow us to distinguish primary tumors from metastases. Only the biopsy can
Fig. 2 – Abdominal ultrasonography: a complex cystic mass on the right ovary, heterogeneous, with both cystic and solid component and some hyperechoic mural nodules and thick septum.

Fig. 3 – Abdominopelvic CT scan: a thickening and an infiltration of the sub-cutaneous space of the umbilical area.
confirm them. In our study, biopsy of the umbilical region revealed umbilical metastasis from adenocarcinoma of gastric origin.

The JMS nodule is most often isolated. Its association with other metastases is rare and unusual, that was the case in our study where it was associated with Krükenberg’s tumor.

Krükenberg’s tumor is an ovarian metastasis which is bilateral in about two-third of cases [10]. It is very rare and represents only 1%-2% of ovarian tumors [3]. Ultrasound and CT scan shows an ovarian mass like a multi-cell solidocystic tumor as in our case. However, imagery does not enable in any way to differentiate the primary ovarian tumor from a secondary tumor [13].

The histologic study is the only one able to confirm the diagnosis. On microscopy, the Krükenberg tumor is characterized by the presence of epitheliomatous cells in a “kitten ring” with an eccentric nucleus filled with muci-carminophilic mucus, isolated or grouped in clusters within fibrils and by a pseudo-sarcomatous proliferation of the stroma. [14]. This histologic aspect in favor of a secondary tumor was also objectified in our case. Primary cancer is in 90% of cases of digestive origin including 70% gastric, 14% colic, 5% pancreatic-biliary, and 2% appendicular, even more rarely mammary and thyroid [15].

The histologic study of gastric buds biopsied after an esogastro-duodenal endoscopy made it possible to make the diagnosis of a well-differentiated gastric adenocarcinoma.

The prognosis of gastric adenocarcinoma is poor and patients are generally non-operable and they are subject only to palliative chemotherapy in the presence of the JMS nodule which is a pejoravite factor [16]. Also, in most of our countries with limited resources, the diagnosis of the primary tumor is late because the management of a JMS nodule is delayed due to its ignorance. Its association with the Krükenberg’s tumor made erring the search for the primary tumor. Failure to achieve immunohistochemistry markers limits us in our therapeutic approach.

It had been instituted in our patient, 6 treatments of chemotherapy but the death occurred 1 month after the beginning of the treatment.

Conclusion

Sister Marie Joseph’s nodule is a rare skin metastasis from an abdominopelvic tumor, most often of gastrointestinal origin. The JMS nodule is most often isolated. Its association with other metastases is rare and unusual such as Krükenberg’s tumor which is a rare ovarian metastasis also secondary to digestive cancer, most often gastric. The presence of this nodule is a pejorative element of the prognosis thus imposing an early and systematic screening of the primary tumor for optimal management which is delayed in most of our countries because of limited resources on the 1 hand, lack of knowledge of the JMS nodule on the other hand and the non-achievement of immuno-histochemical markers limiting us in our therapeutic approach. It had been instituted in our patient, 6 courses of chemotherapy but the death occurred 1 month after the beginning of the treatment.

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