Mesenchymal chondrosarcoma metastasising to the pancreas

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SUMMARY

The mesenchymal chondrosarcoma (MC) is a rare malignant tumour and accounts for less than 3% of primary chondrosarcomas. Mostly MC arises from the craniofacial bones, the ribs, the ilium, the femur and the vertebrae. A 54-year-old man was treated due to an icterus of unknown origin. The medical history of the patient consists of a multimodal treated MC of the thoracic vertebrae. A CT imaging identified a 2×4 cm sized mass of the pancreatic head. Suspecting a pancreatic head carcinoma surgical removal was performed. Histopathological a metastasis of MC was diagnosed. Our patient left the hospital after 17 days and died 23 month after surgery. Metastases of MC to the pancreas are rare. When detecting a mass of the pancreas in patients with a medical history of an MC, a metastasis of these tumour should be taken in consideration.

BACKGROUND

The mesenchymal chondrosarcoma (MC) is a rare malignant tumour and was first described by Lightenstein and Bernstein in 1959.[1] Macroscopically, this rare variant of a chondrosarcoma usually appears as a grey, well-defined, circumscribed mass with a diameter of up to 30 cm.[2] Microscopically, the tumour is characterised by poorly differentiated small round cells and areas of well-differentiated hyaline cartilage.[2] The small round cell components are immunhistochemically positive for SOX9, CD 99 and desmin.[2 3]

CASE PRESENTATION

In February 2017, a 54-year-old man was treated in our hospital due to an icterus of unknown origin.

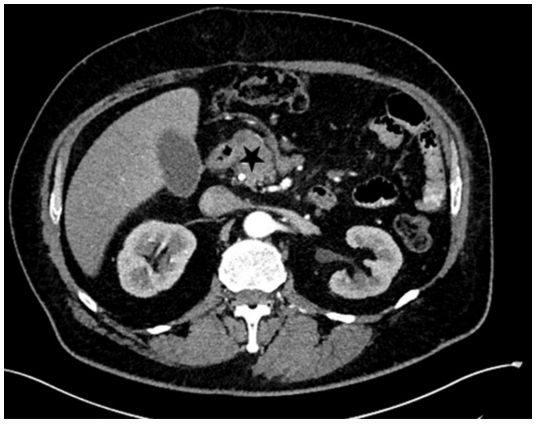

Figure 1 The CT imaging detected a 2×4 cm sized tumour of the pancreas head (marked by black star).

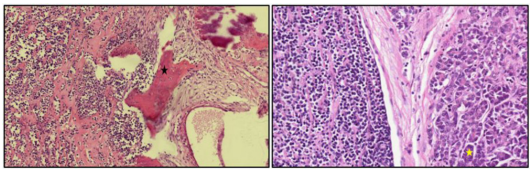

Figure 2 Histopathological examination revealed a mesenchymal chondrosarcoma. The left picture shows an abrupt transition between cartilaginous islands marked by black star and small blue cell components (HE staining, magnification ×10). The right picture shows the small cell proliferation on the left side surrounded by a myopericytomatous pattern in HE staining. The pancreatic tissue is marked by a yellow star (magnification ×20).

His medical history includes multimodal therapy of a rectal carcinoma in 2004. The patient had a palsy of the plexus brachialis with Horner's syndrome following a motorcycle accident. Furthermore, in March 2015, he suffered from an acute spinal ataxia with paraparesis.

DIFFERENTIAL DIAGNOSIS

A MRI revealed a tumour infiltration of the third thoracic vertebrae with compression of the spinal canal. A laminectomy with dorsal spondylodesis and marginal tumour resection was performed. The histopathological examination revealed an MC. The patient subsequently underwent adjuvant radiotherapy, chemotherapy and had no further tumour progress for 12 months.

In December 2016, the patient was referred to our hospital due to an icterus of unknown origin. We performed an endoscopic retrograde cholangiography with implantation of a bile duct-stent due to a stenosis of the ductus choledochus. A CT imaging identified a 2×4 cm sized ill-defined hypodense mass of the pancreatic head (figure 1), an 8 cm sized soft tissue tumour of the left proximal thigh and a disseminated bone metastasis of the sacral bone, the right ilium and the ribs.

Following an interdisciplinary case discussion at our sarcoma board, we suspected a prognosis-determining pancreatic head carcinoma. We assigned the soft tissue tumour of the left proximal thigh and the bone metastasis to the known chondrosarcoma.

TREATMENT

An explorative laparotomy was indicated and performed. Intraoperatively, a tumour of the pancreas head without any evidence of macroscopic peripancreatic infiltration was detected. We performed a pylorus-preserving pancreatic head

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Table 1 Publications on mesenchymal chondrosarcoma (MC) of the pancreas

Author	Year of publication	Gender	Age	Location	Size	Pathological findings	Treatment	Outcome
			Years	Head/corpus/tail	cm	Secondary/primary MC	Month of survival after surgery	
Byun *et al*[15]	1995	Female	36	Tail	7,7×4,3	Secondary	Distal pancreatectomy, CT	NR
Komatsu *et al*[16]	1999	Female	28	Tail	5,5	Secondary	Distal pancreatectomy	NR
Yamamoto *et al*[17]	2001	Male	29	Corpus, Head	NR	Secondary	Enucleation (pancreas head) Distal pancreatectomy	>120
Yamamoto *et al*[17]	2001	Female	40	Head	NR	Secondary	pylorus-preserving pancreaticoduodenectomy	>72
Naumann *et al*[18]	2002	Female	24	NR	NR	Secondary	RT, CT	>84*
Trembath *et al*[19]	2003	Female	27	NR	9,5	Secondary	CT, partial pancreatectomy	NR
Chatzipantelis *et al*[20]	2006	Male	26	Tail	3,8×3,5	Secondary	Distal pancreatectomy	NR
Oh *et al*[21]	2007	Male	41	Corpus, Tail	13×12 x7	Primary	Enucleation	NR
Bu and Dai[22]	2010	Female	34	Corpus, Tail	18×16	Primary	Surgical resection of pancreas body and tail	>52
Tsukamoto *et al*[23]	2014	Male	39	Corpus, Tail	5×6	Secondary	Distal pancreatectomy, CT	34
Guo *et al*[24]	2015	Male	40	Corpus	2×3×2	Secondary	Distal pancreatectomy	>108
Smith *et al*[7]	2015	Female	44	Corpus	NR	Secondary	Distal pancreatectomy, CT	>24
Cohen *et al*[14]	2016	Female	32	Tail	2,9	Secondary	Laparoscopic distal pancreatectomy, CT	15
Cohen *et al*[14]	2016	Female	38	Tail	9,5	Unknown	Distal pancreatectomy, systemic neoadjuvant CT	40

*Month of survival after diagnosis of MC.
CT, chemotherapy; MC, mesenchymal chondrosarcoma; NR, not recorded; RT, radiotherapy.

resection. The postinterventional course was prolonged due to a wound infection and a cholangitis. After wound revision and negative pressure wound therapy, we performed a secondary wound closure. The cholangitis was treated with antibiotics. The patient left the hospital after 17 days of treatment. The histopathological examination diagnosed surprisingly a 40 mm sized metastasis of the known extraskeletal MC of the pancreatic head. A lymphangioinvasion and hemangioinvasion, cone-shaped extension in the ductus choledochus, and another peripancreatic lymph node metastasis in one of 17 resected lymph nodes was revealed (figure 2). Immunohistochemically the tumour expressed S100.

OUTCOME AND FOLLOW-UP

After a case discussion in our sarcoma board we recommended a palliative chemotherapy with epirubicine and ifosfamide. As follow-up MRI 6 month after chemotherapy was advised. The patient died 23 months after surgery.

DISCUSSION

The aetiology of MC is unclear, however, in some cases various abnormalities on chromosome 8 eight have been described in literature.[2 4] MC accounts for less than 3% of primary chondrosarcomas. Men and women are considered to be affected equally. The peak incidence is in the second and third decade of life.[2 5] MC most commonly arises from the craniofacial bones, the ribs, the ilium, the femur and the vertebrae.[5] The primary MC rarely arises from extraskeletal, as the most common extraosseous involvement MC can be determinated in the somatic soft tissue and the meninges.[1] Also, intra-abdominal occurrences of MC are described in literature.[5]

Clinically, patients with MC very often suffer from pain due to the space-occupying effect caused by the growing tumour.[2 6]

The Ewing sarcoma and the enchondroma are described as the main differential diagnosis.[2]

To diagnose MC, a CT or an MRI is advised.[2] The skeletal lesions appear mostly lytic and destructive with poor margins. Nevertheless, in comparison with conventional chondrosarcoma, the radiological features are unspecific. Chondroid-type calcification with contrast medium uptake may be revealed.[2] Moreover, a fine-needle aspiration has been described as a sufficient diagnostic approach.[7]

This tumour entity is very rare and therefore, randomised clinical trials examining the effect of chemotherapy are lacking. A clear treatment strategy has not yet been defined. Generally, the therapy of MC consists of radical surgery and chemotherapy (methotrexate, doxorubicin, cisplatin and ifosfamide). But also neoadjuvant therapy in a non-metastatic stage and radiotherapy as a therapeutic approach have been published.[6] After interdisciplinary case discussion at our sarcoma board an exploratory laparotomy was indicated. Not suspecting a metastasis of MC we made this decision. And to prevent local complications we did not conduct a fine needle aspiration according to our German guidelines.[8] With the knowledge of a possible survival for years even in a metastatic stage, we wanted to surgical treat the suspected pancreas carcinoma.

The prognosis is poor and distant metastasis may occur much later on. The majority of metastasis of MC is skeletal. But also the intra-abdominal appearance of MC like in the case report at hand has been described in literature. To that some case reports were published concerning the primary and secondary occurrence of MC in the kidney as a rare extraperitoneal manifestation.[9 10] Moreover, Suzuki *et al* treated a patient suffering from

MC of the uterus.[11] To our knowledge, only one case report of MC in the spleen was described in literature.[12]

Secondary tumours of the pancreas have been published to compose approximately 4% of pancreatic masses.[13] Metastasis of MC to the pancreas is rare. Local recurrence may occur.[14] We reviewed the literature using Pubmed and Google Scholar (table 1). The search yielded 12 relevant publications containing 14 cases (nine women, five men) of MC of the pancreas.[7 14–24] The mean age was 34 years (range, 24–44). In most of the cases, the pancreas tail was affected (n=7). Only one patient suffered from MC of the pancreas head such as in the case report at hand. In two cases, MC primary appeared and in 11 cases, MC appeared secondary in the pancreas. With one exception MCs were surgical resected. Six patients underwent pancreatic surgery and had metastases of other sites. Most likely these authors did not expect a metastasis of MC either. The survival after surgery ranged in these cases between 15 and 120 months.[14 17 19 20 24] To that Schneiderman *et al* conducted a survival analysis of MCs. Describing metastases as independent predictor of death a 44% 10-year-overall survival rate of patients suffering from extraskeletal MC has been described.[25]

Learning points

- Metastases of mesenchymal chondrosarcoma to the pancreas are rare. To our knowledge only 14 cases are published.
- When detecting a mass of the pancreas in patients with medical history of MC, a metastasis of this tumour entity should be taken in consideration.

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