Case Report

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Pancreatic metastasis of mesenchymal chondrosarcoma: a surgical case report and review of literature

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Introduction: Mesenchymal chondrosarcoma (MC) is a rapidly progressive sarcoma that predominantly impacts the bones. Making up only 3% of chondrosarcomas, about one-third of these tumours develop in extra-skeletal sites.
Case presentation: The authors present a clinical case of a 42-year-old patient who was diagnosed with MC 8 years ago, now admitted to the hospital with a palpable epigastric mass. Clinical and laboratory examinations showed consistent results for MC tumours, with metastasis to the body and tail of the pancreas and invasion of the splenic vein. Surgical resection and systemic screening were performed to ensure that there were no lesions elsewhere. Regular follow-up has found no localized lesions or complications after 15 months.
Clinical discussion: Metastatic extra-skeletal mesenchymal chondrosarcoma of the pancreas is exceptionally rare. To our current understanding, only 14 such cases have been documented in medical literature. The symptoms of pancreatic metastasis are diverse and the radiographic features of metastatic mesenchymal chondrosarcoma are not typically distinct.
Conclusions: Although MC tumours do not frequently occur in sites other than the axial system, a tumour presenting later in a patient with a history of MC should be reviewed to confirm the diagnosis of metastatic MC. Treatment can vary between surgery, radiation therapy and systemic therapy.

Keywords: case report, mesenchymal chondrosarcoma, pancreatic metastasis

Introduction

In 1959, Lightenstein and Bernstein were the first ones to provide the first sketches of Mesenchymal chondrosarcoma (MC)[1]. MC affects both men and women equally, the peak incidence peaks in the second and third decades of life. Characterized by a high histological grade, less than 3% of primary chondrosarcoma are composed of this tumour form, which typically originates from skeletal locations, most commonly the ribs, ileum, femur, and vertebrae[2]. Tumours are connected to incredibly uncommon examples of solid organs[3]. There have been a few reports of irregular extra-skeletal mesenchymal chondrosarcoma (ESMCs), mainly found in the meninges, neck, thorax and intra-abdominal cavity. We report a case of MC with remote metastases to the lung, the body and the tail of the pancreas, due to the rarity of the disease and a shortage of documented cases. Nevertheless, despite the lengthy clinical course, it is characterized by a high rate of metastases, local recurrence as well as high mortality[4].

We describe a clinical case of a male patient with MC disease who developed pancreatic metastases invading the splenic vein 8 years after initial diagnosis. All our work has been reported in line with the SCARE criteria and guidelines[5].

HIGHLIGHTS

- Metastatic extra-skeletal mesenchymal chondrosarcoma is exceptionally rare.
- The clinical manifestations of pancreatic metastasis are variable.
- The diagnosis of pancreatic metastasis may be very challenging and immunohistochemistry staining is mandatory.
- Radical surgery must be still the most effective treatment approach.

Case presentation

A 42-year-old male patient presented to the hospital in March 2015 with a right humerus supracondylar fracture because of an

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unintentional fall while performing routine daily tasks. On admission, the patient was diagnosed with a pathological fracture, since the X-ray showed that the patient had a fracture above the osteoma. Biopsy results showed homology consistent with mesenchymal chondrosarcoma. The patient then underwent interscapular–thoracic amputation. At the 6th follow-up month, the patient was presented with a mass in the lower lobe of the left lung.

A thoracic multidetector computed tomography (CT) scan in 2015 revealed a calcified lobulated mass in the left lower lobe of the lung, which was confirmed metastatic MC on histology (Fig. 1A, B) (white arrow).

The biopsy of the lung mass showed that the mass was an MC tumour that had metastasized to the lung. During the period from December 2015 to May 2016, he received six rounds of the AP chemotherapy regimen (Doxorubicin 60 mg/m^2 at day 1 and Cisplatin 80–100 mg/m^2 from day 1 to day 3 within a 21 days cycle). The Multidisciplinary Tumour Board concluded the indication for left inferior pulmonary lobectomy, yet the patient refused. Patients were evaluated for response to chemotherapy and follow-up was performed every 6 months. No abnormalities were detected until May 2022. The lesion is now completed calcified and does not change size in X-ray and CT scan until now.

About 8 years after the patient's initial treatment, in May 2022, at a post-treatment check-up, ultrasound scans revealed the presence of a mass in the body and tail of the pancreas. An abdominal multidetector computed tomography scan (MCTS) demonstrated a poorly marginated, cystic mass measuring 4.5 × 3.5 cm in the pancreatic body and tail with peripheral calcification (Fig. 2A, white arrow); enhancing capsule and internal bulkheads on arterial phase (B). The tumour infiltrates adjacent peripancreatic adjacent fat, involving the splenic vein (C). Upper abdominal magnetic resonance imaging (MRI) shows an ill - defined cystic lesion of the pancreatic body and tail measuring ~4.5 × 3.5 cm (Fig. 3) with a thick irregular wall and some internal septations that had intermediate signal on T1W (A) and T2W (E), enhancement on post- contrast arterial phase (B) and venous phase (C, D). The intra-cystic fluid had an intermediate signal on T1W (A), heterogeneous T2W hyperintense (E). The upstream pancreatic duct was dilated (Fig. 4).

Peripancreatic infiltration and multiple lymph nodes with intermediate signals on T1W, T2W (not shown).

The patient received a subsequent laparoscopic spleno-pancreatectomy for the rare presentation and tissue proof. The surgical pathology result confirmed the metastatic mesenchymal chondrosarcoma of the body of the pancreas (Fig. 5).

The results of the multidisciplinary consultation were concluded to be consistent with a metastatic lesion secondary to MC.

After surgery, the patient stabilized and was discharged after 15 days. The patient was screened for the whole body in June 2022 to look for a secondary lesion other than the known lesion by CT scan. An abdominal multidetector CT scan 1.5 months

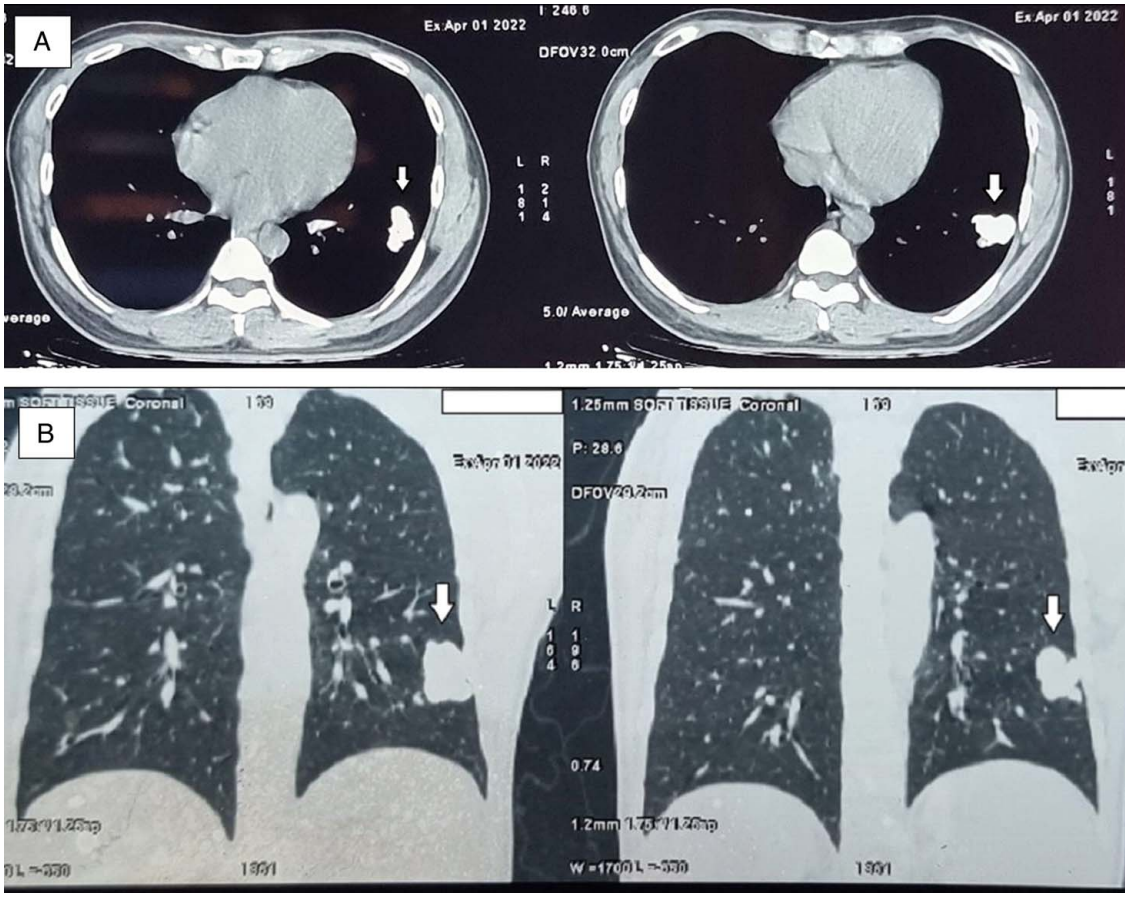

Figure 1. Axial mediastinal window (A) and coronal lung window (B) non-contrast enhanced computed tomography image.

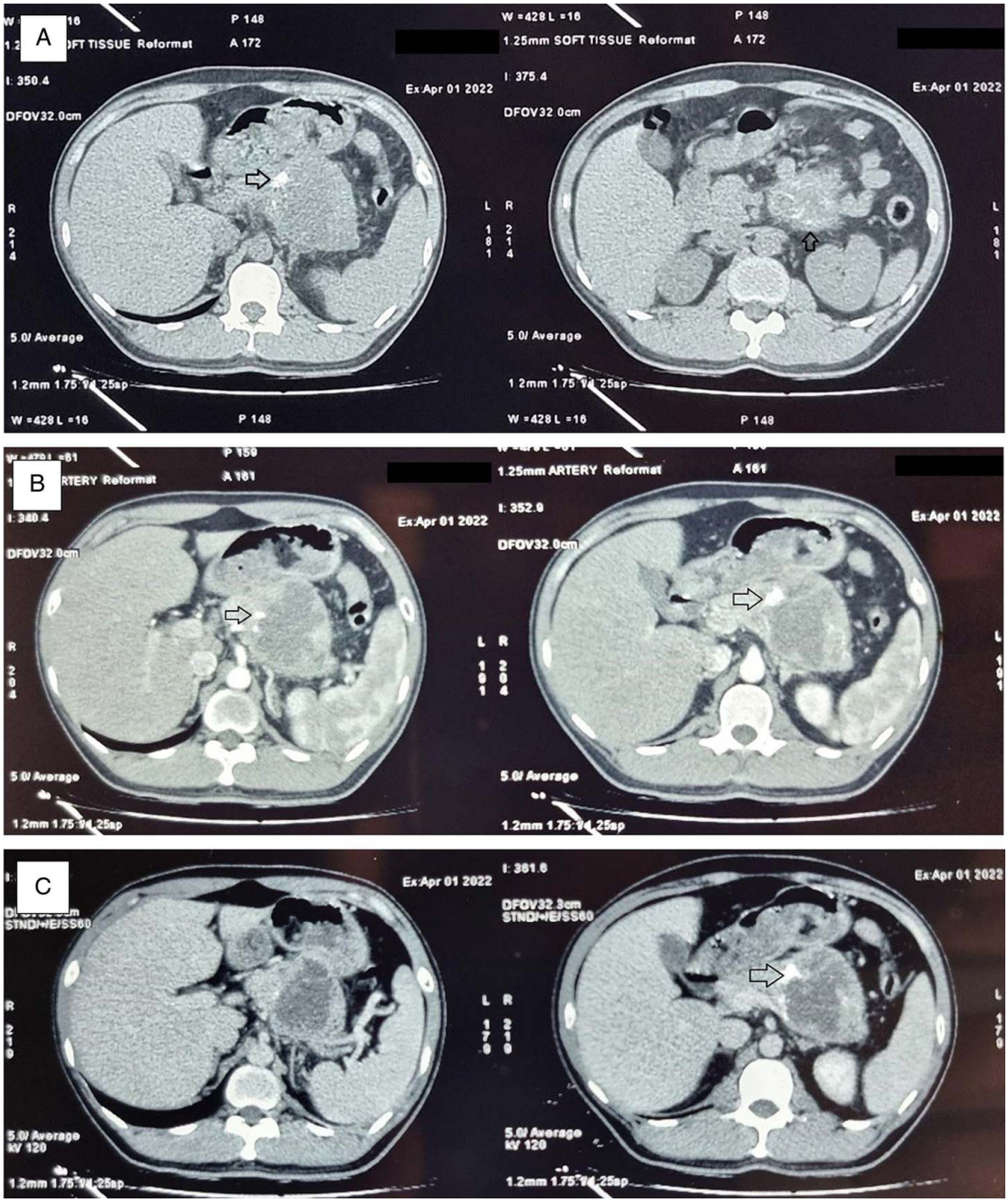

Figure 2. Axial pre-contrast (A) and post-contrast arterial phase (B) and venous phase (C) computed tomography.

after surgery (Fig. 6) showed homogenous pancreatic head tissue with an adjacent suture line (A, B). The pancreatic head was enhanced homogeneously (A, B) and no localized lesions or complications were found. Currently, patients are still monitored every 6 months to check for new abnormalities, if any.

Discussion

Mesenchymal chondrosarcoma is a highly malignant tumour that mainly affects people aged 25–30[6,7]. The pathogenesis of the tumour is currently poorly understood. Contrary to typical chondrosarcoma, mesenchymal tumours most frequently affect the axial skeleton, including the vertebra, ribs, ilium, and craniofacial bones (particularly the jaw)[8,9]. Their developed cartilage is combined with solid, highly cellular regions made of undifferentiated small round cells, which is how they are distinguished. Several observational studies have suggested that the disease is linked to chromosome 8[7].

At 5 and 10 years, the mesenchymal chondrosarcoma overall survival rate is 51% and 43%, respectively[2]. There is no distinction in overall survival between extra-skeletal and skeletal

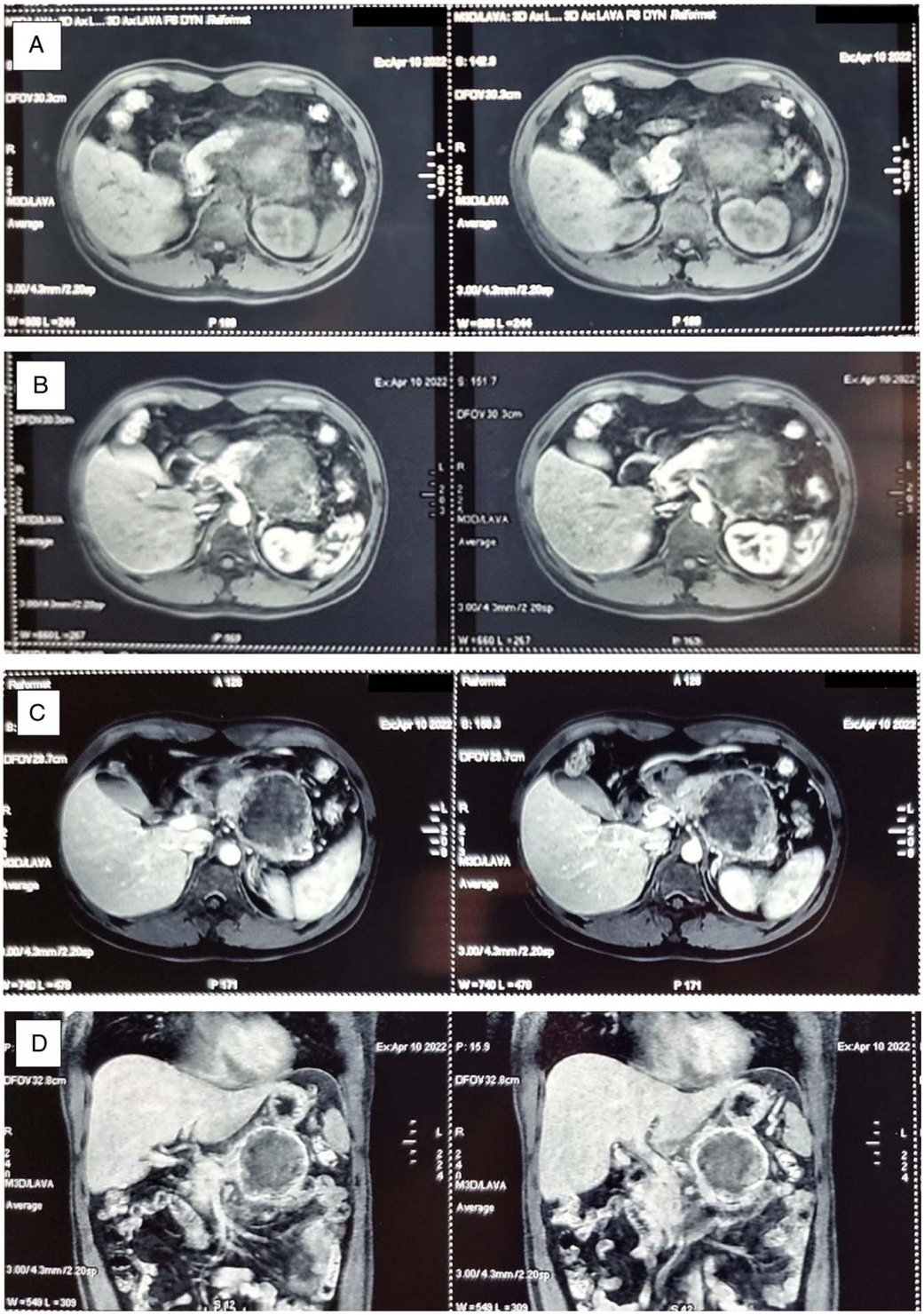

Figure 3. Axial pre-contrast (A) and post-contrast arterial phases (B) and axial (C), coronal (D) venous phases T1-weighted fat-suppressed; axial T2-weighted fat-suppressed (E). Further non-invasive and invasive tests show: a left-sided pancreatic tumour invading the splenic vein.

cancers. Both the existence of tumour metastasis and an increase in tumour size of 1 cm were independently linked to a higher mortality risk[10]. Given this, mesenchymal chondrosarcoma patients typically have poor prognoses and require ongoing monitoring.

The symptoms of pancreatic metastasis are diverse; some symptomatic patients may experience abdominal pain and jaundice; however, most patients do not suffer from organ-specific pain, and the metastasis is only discovered during normal follow-up[11]. Therefore, the early detection of secondary pancreatic tumours on a

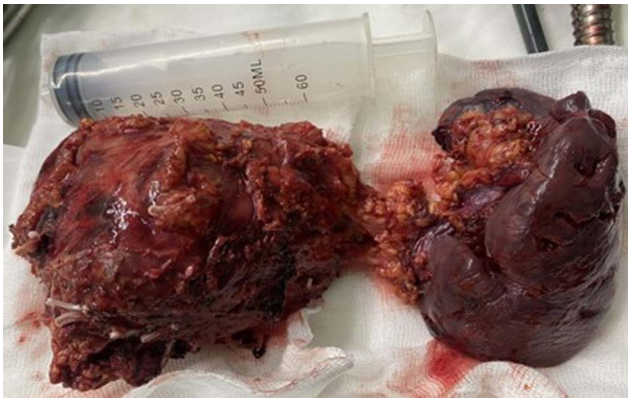

Figure 4. The specimen is extracted by a mini laparotomy in the middle line.

CT scan and MRI is critical[12]. Mesenchymal chondrosarcoma has a less defined appearance than conventional chondrosarcoma. To diagnose pancreatic masses, fine-needle aspiration of the pancreas has become increasingly utilized[11].

The radiographic features of metastatic mesenchymal chondrosarcoma are not typically distinct. In the case we observed, the pancreatic tumour exhibited some peripheral calcification and a central fluid necrosis. These characteristics can also be seen in various other conditions, including cystic pancreatic neoplasms (like mucinous cystadenoma or carcinoma, solid pseudopapillary tumours, or pancreatic neuroendocrine tumours). Moreover, the patient was found to have a calcified lesion in the left lung, which was later confirmed as a metastasis. This pointed towards a probable pancreatic metastasis prior to the surgery. Consequently, we performed a distal partial pancreatectomy to make a definitive diagnosis and establish the most appropriate therapeutic approach, inclusive of chemotherapy.

Metastatic extra-skeletal mesenchymal chondrosarcoma (EMC) of the pancreas is exceptionally rare. To our current understanding, only 14 such cases have been documented in medical literature (Table 1). Therefore, our case should be considered the 15th reported instance of pancreatic EMC in the literature. The average age was 35, with no discernible gender difference, comprising 6 males out of the total 14. The average largest dimension of the tumour was 4.7 cm, with the majority being in the body and tail of the pancreas in 11 out of 14 cases. Of all the reported cases, calcification within the pancreatic tumour was noted in six cases. The average latency period for pancreatic metastasis following the detection of the primary tumour was approximately six years.

There are no specific recommendations for the treatment of this type of tumour; of the reported cases, most treatment strategies were surgery with further systemic chemotherapy (Table 1). According to the outcomes so far, either a chemotherapy plan based on osteosarcoma-type doxorubicin plus cisplatin, or an Ewing sarcoma-based multidrug regimen may be used. If the tumour is at the non-metastatic stage, neoadjuvant therapy may be considered[23,24]. Additionally, radiation as a treatment strategy has been documented.

In the presented case, an unusual presentation with an asymptomatic splenic invasion of the pancreatic tail was the second distant metastasis detected eight years after the initial diagnosis. We arranged a spleno-pancreatectomy for surgical resection. Histological examination confirmed a metastatic mesenchymal chondrosarcoma. This demonstrates the importance of long-term follow-up after successful treatment.

Conclusion

Mesenchymal chondrosarcoma is a rarely seen tumour with a dismal prognosis. The patient's prognosis for survival is adversely impacted by the size of the tumour and the number of metastatic locations. Skeletal locations are susceptible to tumour development; however, extra-skeletal areas require caution. Any site where abnormal masses appear, especially with calcification inside on imaging, should be guarded against the appearance of secondary lesions. Long-term follow-up after treatment, as well as systemic screening, is important in

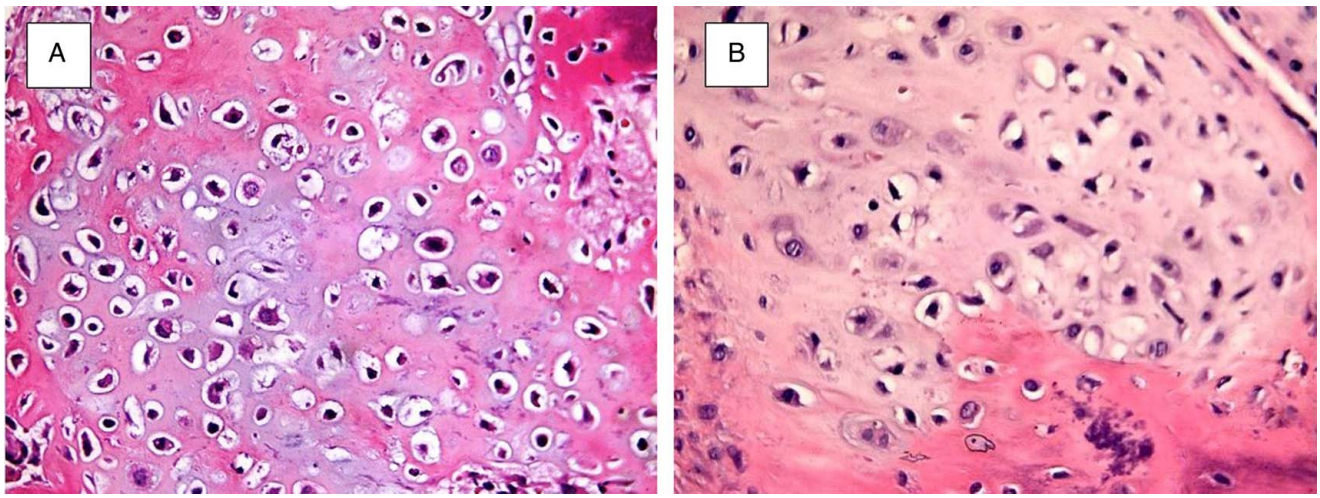

Figure 5. (A) Immunohistochemical staining of the pancreatic mass was performed. The histological analysis demonstrated the presence of mesenchymal cells that exhibited a tiny, round, and spindle-shaped morphology (blue arrow, magnification 1000×). (B) Furthermore, the image depicts the presence of distinct cartilage islets (denoted by a white star) that exhibit a sudden transition from the surrounding blue spherical cells (B, 1000×).

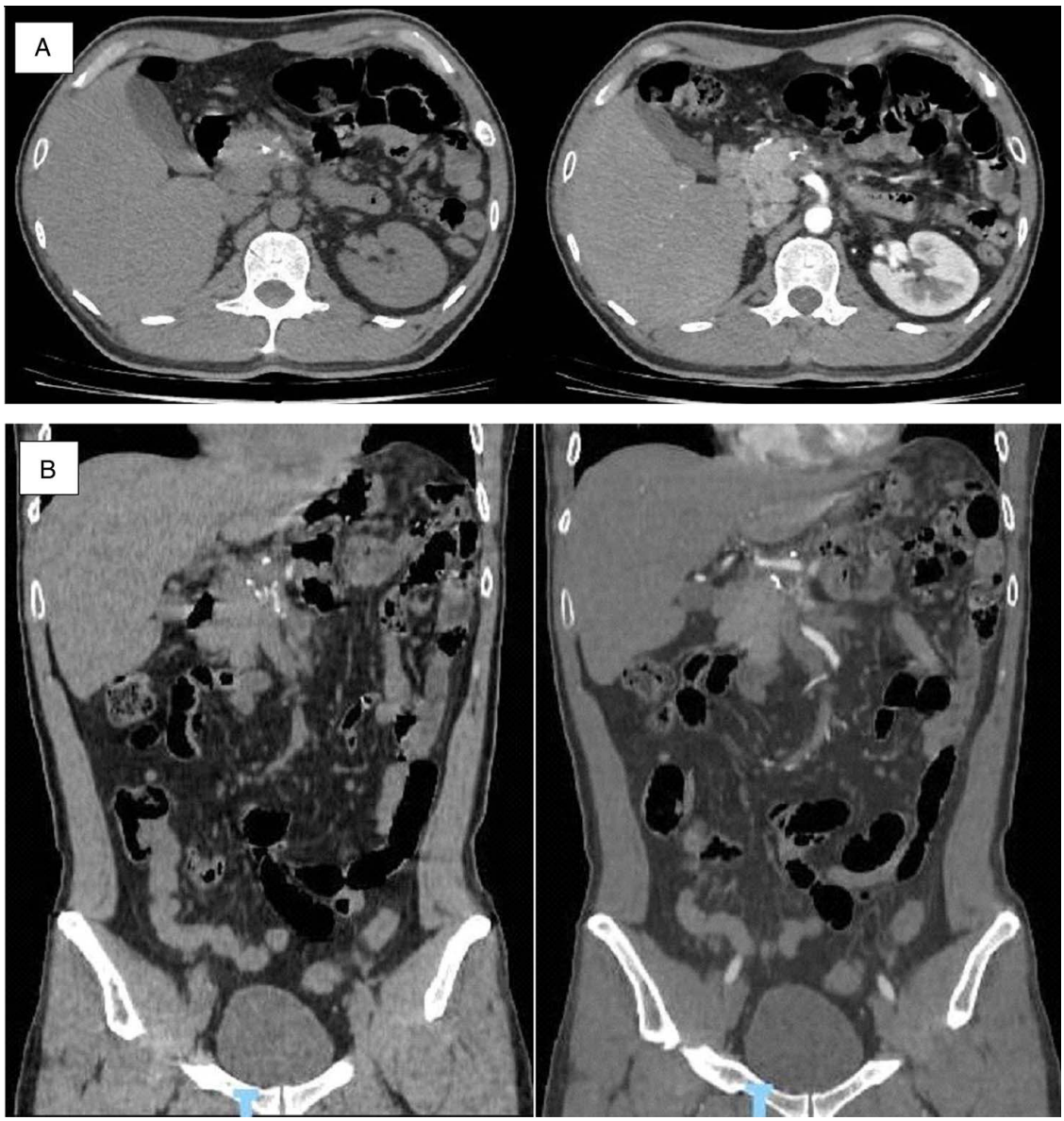

Figure 6. Axial (A), coronal (B) abdominal computed tomography pre-contrast (left) and post-contrast (right) arterial phase.

the MC situation. The diagnosis should be a combination of multidisciplinary and skilled physicians.

Ethics approval

The study was approved by the Research Ethics Committee of Hue Central Hospital. The procedures used in this study adhere to the tenets of the Declarations of Helsinki.

Consent

Written informed consent was obtained from the patient for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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The authors declare no funding for this study.

Authors contribution

H.N.D.: conceived, performed the operation and wrote the manuscript. P.A. T., T.N.D., and T.T.L.: performed the operation and edited the manuscript. V.T.T.L., H.H.T. N., and H.T. L.: edited the manuscript. All authors contributed to the interpretation of the results, discussed the results. All authors read and approved the final manuscript to submit.

Conflicts of interest disclosure

The authors declare that they have no conflicts of interests.

Table 1

Literature review of a published case of the metastatic mesenchymal chondrosarcoma of the pancreas

References	Year	Sexr	Age	Size (cm)	Location (head, body, tail)	Primary site	Metastatic site	Latency period for pancreas metastasis	Pancreatic tumour with calcified deposit	Treatment
Byun *et al.*[13].	1995	Female	36	7.7 × 4.3 × 5	Tail	Thigh	Pancreas	Synchronous	NR	Distal pancreatectomy, CT
Komatsu *et al.*[14]	1999	Female	28	2.5	Tail	Meninges	Pancreas	17	Yes	Distal pancreatectomy
Yamamoto *et al.*[15]	2001	Male	29	NR	Body, Tail	Thigh	Pancreas, lung, testis, skin, chest wall	3	Yes	Distal pancreatectomy
Naumann *et al.*[16]	2002	Female	24	NR	NR	Retroperitoneum	Kidney, lung, rib, humerus, pancreas, spine	6	NR	RT, CT
Trembath *et al.*[17]	2003	Female	27	9.5	NR	Tibial	Retroperitoneum, pancreas, diaphragm	2	NR	Partial pancreatectomy, CT
Chatzipantelis *et al.*[18]	2006	Female	26	3.8 × 3.5	Tail	Brain	Lung, thigh, pancreas	9	NR	Distal pancreatectomy
Tsukamoto *et al.*[4]	2014	Male	39	5 × 6	Body, tail	Buttocks	Pancreas, sacrum, ilium, ischium, lungs	Synchronous	NR	Distal pancreatectomy, CT
Smith *et al.*[11]	2015	Female	44	NR	Body	Chest wall	Pancreas	21	NR	Distal pancreatectomy, CT
Guo *et al.*[19]	2015	Male	40	2 × 3 × 2	Body	Femoral vein	Pancreas, lung, pleural, mediastinal and axillary node	3	Yes	Distal pancreatectomy
Cohen *et al.*[20]	2016	Female	32	2.9	Tail	Pterygoid region	Pancreas, lung	8	Yes	Laparoscopic distal pancreatectomy, CT
Shah *et al.*[12]	2019	Male	49	5	Tail	Thigh	Pancreas	10	Yes	Distal pancreatectomy
Camacho *et al.*[21]	2020	Female	53	3	Tail	Lower limb	Lung, pancreas	7	NR	Laparoscopic distal pancreatectomy
Sun *et al.*[22]	2020	Male	21	4.94	Neck	Rib	Pancreas, adrenal gland	Synchronous	NR	NR
Chen *et al.*[5]	2021	Male	34	3.4	Tail	Chest wall	Pancreas	1	Yes	Distal pancreatectomy, CT
Present case	2022	Male	42	4.5 × 3.5	Body, tail	Humerus	Lung, Pancreas	8	Yes	Laparoscopic distal pancreatectomy, CT

The table originates from Chen *et al.*[5], with a new column (highlighted) added by this report.
CT, chemotherapy; NR, not recorded; RT, radiotherapy.

Research registration unique identifying number (UIN)

HCH.2904.

Guarantor

Hung N. Dang.

Availability of data and material

Data are available upon reasonable request and with permission of Hue Central Hospital.

Provenance and peer review

Not commissioned, externally peer-reviewed.

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