



## Case Report

# A case of an upper anterior abdominal wall extra-skeletal Ewing sarcoma-soft tissue reconstruction with medial intercostal artery perforator flap and free anterolateral thigh fasciocutaneous flap with arteriovenous loop graft

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## ABSTRACT

**Introduction:** Extra-skeletal Ewing sarcoma - a rare clinical entity. After the tumour resection, a huge upper anterior abdominal wall defect poses a challenge in soft tissue reconstruction in a thin individual.

**Case presentation:** We report an anterior abdominal wall Ewing sarcoma in a 22-year-old gentleman, arising from the left rectus abdominis muscle. After wide local excision, the reconstruction was achieved with free anterolateral thigh (ALT) fasciocutaneous flap with arteriovenous (AV) loop graft, right medial intercostal artery perforator (MICAP) flap and split-thickness skin graft (SSG).

**Discussion:** The goals of anterior abdominal wall reconstruction are to restore the integrity of the abdominal wall, prevent visceral eventration, and provide functional support. In view of the size of the defect, a free ALT flap was harvested and anastomosed to the left deep inferior epigastric bundle with the AV loop graft. Although the current trend is skewed towards the use of biologic mesh, an on-lay prolene mesh was used due to its affordable cost and that the biologic mesh was not available in Malaysia. The options of further reconstruction after the patient developed marginal flap necrosis and surgical site infection were also discussed. Post-operation 3 months, there was denser hair growth on the ALT flap. This finding has never been reported before and warrants further studies.

**Conclusion:** The use of combination of various technique, namely free ALT fasciocutaneous flap with AV loop graft, right MICAP flap and SSG in reconstruction ensures a satisfactory functional and aesthetic outcome in the upper anterior abdominal wall reconstruction.

## 1. Introduction

Extra-skeletal Ewing sarcoma (EES) is a type of very uncommon soft tissue sarcoma. Here, we present a case of reconstruction of an extensive upper anterior abdominal wall defect, after wide local excision of the EES in a thin gentleman. As a result of its versatile and robust nature, anterolateral thigh flap has been used extensively for various post-oncological resection defects in the abdominal region. We managed to achieve a satisfactory closure with free ALT fasciocutaneous flap with AV loop graft, as well as right medial intercostal artery perforator flap and split-thickness skin graft after multiple wound debridement due to surgical site infection.

## 2. Case presentation

A 22-year-old gentleman, previously had no known medical illness, presented with a rapidly growing anterior abdominal wall mass over a period of 5 months. Prior to this, he had sustained a blunt trauma to that region. He did not have any other constitutional signs or symptoms and his blood parameters were all within normal range. On examination, an ulcerating, fungating mass measuring 15cm × 15cm was noted, with its base measuring 9cm × 9cm on the left upper quadrant of the abdomen (Fig. 1a and b). The computed tomography (CT) and magnetic resonance (MR) images of the abdomen revealed that the tumour was likely a pedunculated soft tissue sarcoma arising from the left rectus abdominis muscle (Fig. 2a and b).

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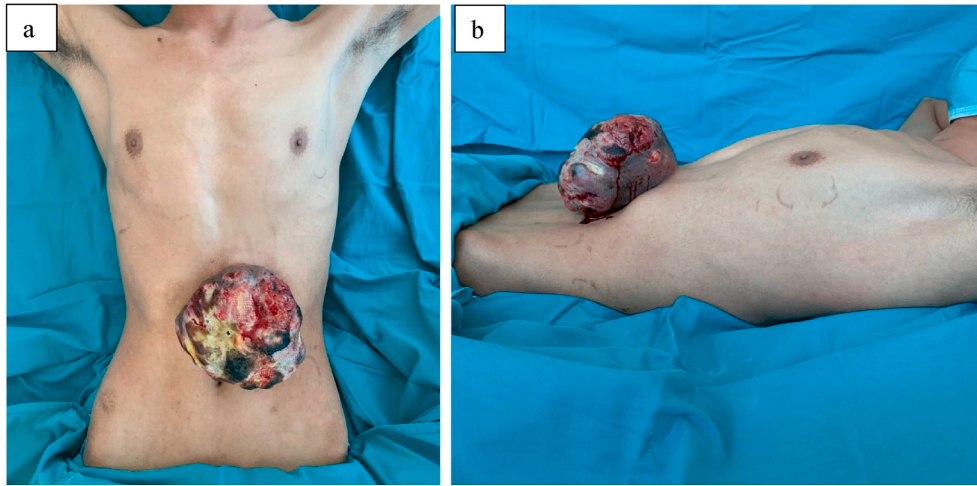
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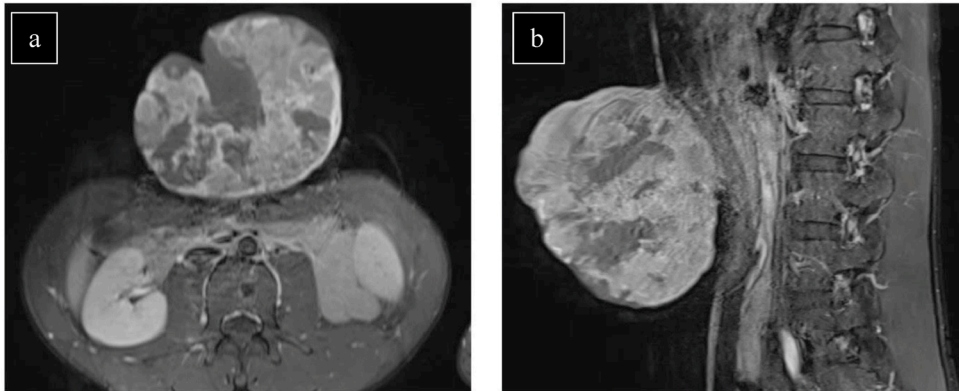
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**Fig. 1.** a(anterior view), b(left lateral view)- An ulcerating, fungating mass with bleeding tendency, measuring 15cm × 15cm with its base measuring 9cm × 9 cm on the left upper quadrant of the abdomen.

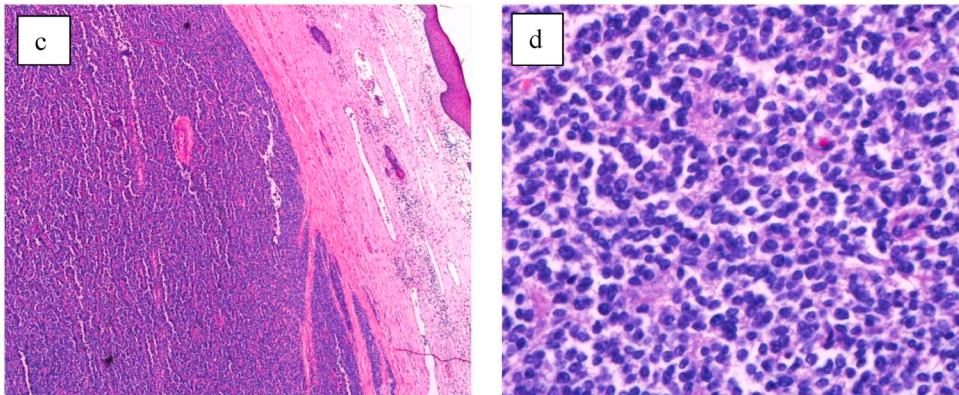


**Fig. 2.** a(transverse view), b(sagittal view)- MR abdomen demonstrated that the tumour was most likely a pedunculated soft tissue sarcoma arising from the left rectus abdominis muscle.

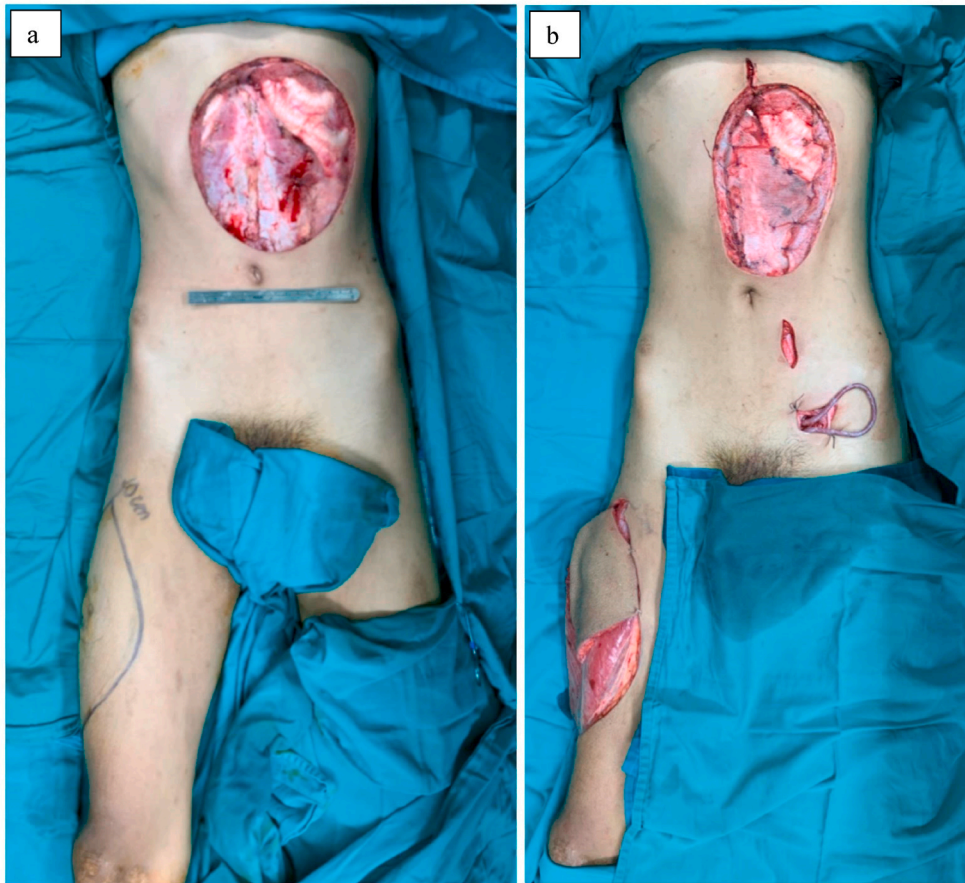
**Fig. 2c-** (Original magnification X4) Sections of tumour show sheets of small round blue cells, in areas separated by fibrous septae. Extensive necrosis are seen.

**Fig. 2d-** (Original magnification X40) The tumour cells exhibit uniform cells with hyperchromatic nuclei with vesicular nuclei with inconspicuous nucleoli and indistinct eosinophilic to clear cytoplasm.

**Fig. 2e-** Excised tumour weighing 1kg. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)







**Fig. 3.** a- Intraoperatively, the post-excision anterior abdominal wall defect was measured 20cm × 16cm (CC x W) and the posterior rectus sheath was left intact.

**Fig. 3b-** The defect was subsequently downsized and an on-lay prolene mesh was applied. Right free ALT fasciocutaneous flap was raised to fit into the defect with the creation of AV loop grafted from the right long saphenous vein. The AV loop graft can be seen over the left iliac fossa after anastomosing with the deep inferior epigastric vessels (recipient vessels).

Wide local excision with 3cm margin was performed. The post-excision defect rendered at anterior abdominal wall was measured 20cm × 16cm (CC x W) and the posterior rectus sheath was left intact (Fig. 3a). The defect was subsequently downsized with undermining of wound edges, and an on-lay prolene mesh was applied. Anterolateral thigh (ALT) fasciocutaneous flap was raised from the right thigh to obliterate the defect, with the creation of arteriovenous (AV) loop graft procured from the right long saphenous vein (Fig. 3b) to facilitate microvascular anastomoses to deep inferior epigastric artery and vein.

The histopathology study showed small round blue cell tumour, consistent with Ewing sarcoma (Fig. 2c and d). All resected margins were free from the tumour. Post-operatively, the patient experienced marginal flap necrosis and surgical site infection (Fig. 4b). The infected mesh was partially removed and multiple wound debridement were required to obtain satisfactory wound bed. Eventually he underwent closure with right medial intercostal artery perforator (MICAP) flap and split-thickness skin graft (SSG) over the remaining defect (Fig. 4c, d, e, f, g, h).

The patient was discharged home with no further wound complications. Post-operative CT of the thorax, abdomen and pelvis revealed no distant metastasis and the patient was planned for chemoradiotherapy by the oncological team. Upon review in out-patient clinic 3 months post-operatively, there was denser hair growth noted over the ALT flap (Fig. 5). The patient also noted that he required his upper limbs' assistance in the forward flexion of the trunk. Otherwise, he was able to ambulate independently and has not shown any signs of abdominal hernia.

### 3. Discussion

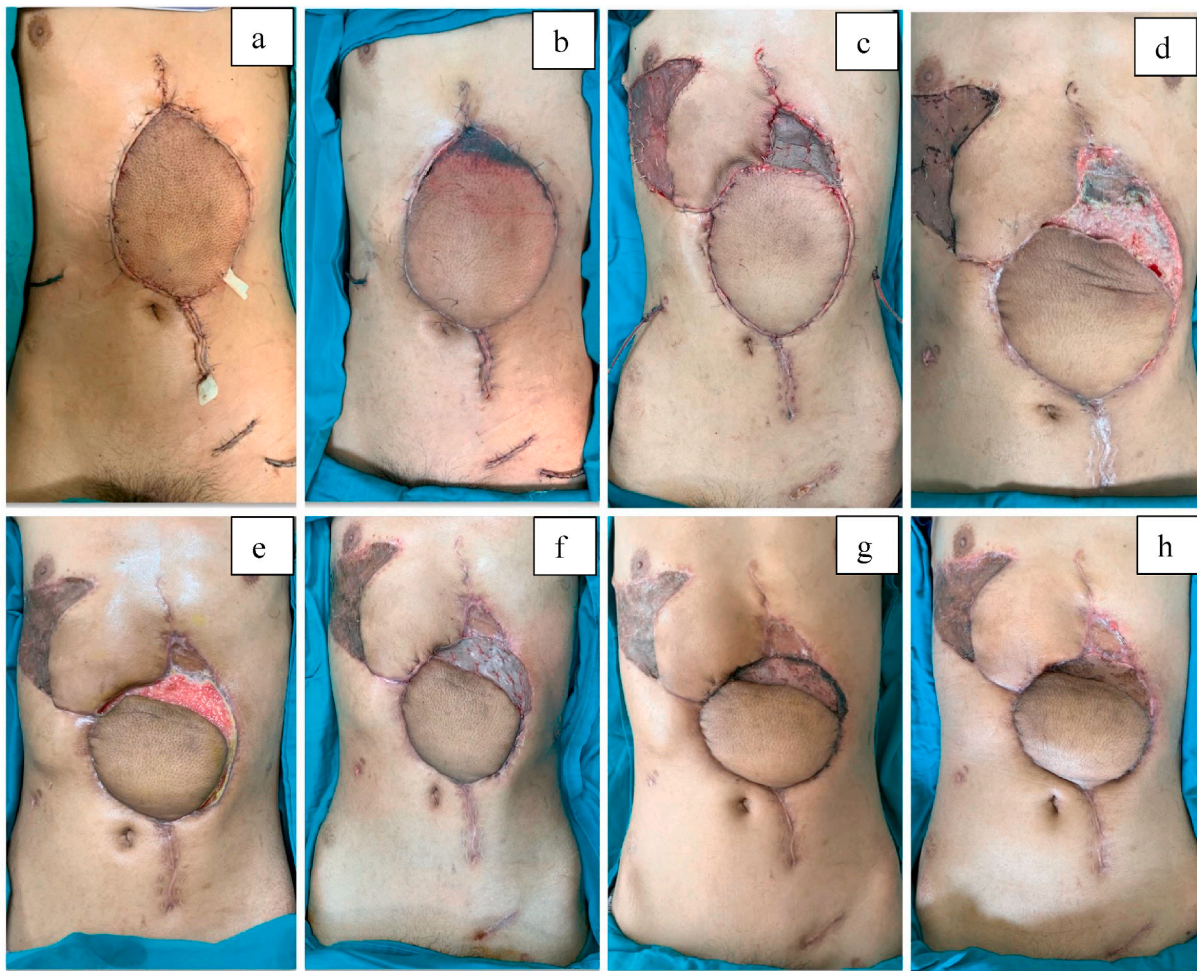
Ewing sarcoma is known to be the second most common primary bone sarcoma affecting children and adolescents after osteosarcoma [1].

It can also occur exclusively in the soft tissue, which is classified as extra-skeletal Ewing sarcoma (EES) [2]. It is more common in male and has an affection towards children, adolescents and young adults (AYAS) with around 1.5 cases per million children and AYAs globally [2]. EES in particular is more predominant in adults and usually found in the trunk, which corresponds to our patient's presentation.

Ewing sarcoma has an aggressive behaviour, thus approximately 20–25% of patients present with metastasis at the time of diagnosis, and are usually resistant to intensive therapy [2]. EES typically has a better prognosis than skeletal Ewing sarcoma [3]. Our patient did not have any constitutional symptoms besides the growth on the anterior abdomen. In view of the nature of the EES to mimic other soft tissue tumour, the diagnosis was only made post-operatively following histopathological examination. Regardless of the primary site of involvement, Ewing sarcoma is known to be comprised of the characteristic small round blue cells [3].

CT staging was done for the index patient post-operatively to determine the extent of the disease, to identify the possibility of residual tumour and for accurate staging of the disease prior to the oncological intervention. Treatment of Ewing's sarcoma is multimodal, comprising of surgery, radiation, and multidrug chemotherapy [1]. However, EES is a relatively rare entity and there is limited data to guide local therapy [3]. Some studies demonstrate that combined-modality of local therapy with both surgery and radiation therapy are not guaranteed to lower recurrence rate, thus suggesting that the single-modality local therapy alone may be sufficient for select patients [3]. Radiotherapy is indicated in cases where there is residual disease after surgical intervention, or where surgery is contraindicated [3].

The abdominal wall is comprised of a laterally-based musculoaponeurotic system in conjunction with the centrally placed pyramidalis muscles and rectus abdominis with the rectus sheath [4]. The



**Fig. 4.** a- Post-operation day 1, free ALT fasciocutaneous flap was supple and viable. Noted 2 corrugated drains inserted.

**Fig. 4b-** Post-operation day 5, patient developed surgical site infection. A well demarcated marginal necrosis also noted over the cranial part of the free ALT fasciocutaneous flap, which was the distal end from the flap pedicle.

**Fig. 4c-** Post-operation 2 weeks from the first reconstruction, we went in for partial mesh removal and marginal flap debridement, rendering a defect over the cranial part of the wound, which was subsequently covered with right MICAP flap. SSG was then applied to the remaining raw area on the thoracic cage, including the donor site of the right MICAP flap.

**Fig. 4d, e-** Unfortunately, the patient developed surgical site infection again which required multiple bedside wound debridement until healthy granulation tissue was seen. There was only around 50% of SSG taken.

**Fig. 4f-** Post-operation 6 weeks from the first reconstruction, a final wound debridement was carried out and SSG reapplied over the defect of the cranial part of the wound.

**Fig. 4g, h-** Post-operation 7 weeks and 8 weeks respectively from the first reconstruction, the SSG has taken fully and the wound has healed completely.

anterior abdominal wall is crucial in the protection of viscera, postural stabilization, and maintenance of intraabdominal pressure [4]. Therefore, a defect in the anterior abdominal wall can significantly affect these functions. Our patient had an upper abdominal wall defect, measuring 20cm × 16cm (CC x W) after the resection of tumour, which required reconstruction in order to prevent complications such as ventral hernia. It posed a challenge as our patient was very underweight (BMI 15.9kg/m<sup>2</sup>), resulting in insufficient local tissue to manipulate for local tissue transfer. Furthermore, a large upper abdominal defect usually poses great challenge in reconstruction with locoregional flaps. Thus, we decided a free ALT fasciocutaneous flap may solve the reconstructive dilemma. An ALT flap was chosen in view of its long consistent pedicle, large skin coverage, tough fascial supportive layer and our surgeon's preference. A myocutaneous flap was not utilised as the posterior rectus sheath was left in situ to provide a dynamic muscle support. The patient might not have experienced the weakness in forward flexion of the trunk if a functional free tissue transfer such as an innervated chimeric ALT, rectus femoris, tensor fasciae latae flap was used.

Intra-operatively, we encountered another hurdle of looking for eligible recipient vessels because both superior epigastric arteries were not patent. Eventually the left deep inferior epigastric bundle was chosen with the arteriovenous (AV) loop recruited from the right long saphenous vein in view of the insufficient pedicle length of the free ALT flap. The AV loop graft was used in order to achieve a tension-free microvascular anastomosis [5]. Nonetheless, this increased the microvascular complexity and the chance of vessel thrombosis [5].

Mesh reinforcement with overlying myofascial coverage is preferred as a first-line option in anterior abdominal wall reconstruction [4]. The decision to place biologic or synthetic mesh depends on availability, patient's comorbidities, and clinical factors of the case [4]. The ideal mesh should be biocompatible, nonallergenic, non-resorbable, cost-effective, resilient to infection, high tensile strength, radiolucent, and accessible for repeat operative access [6]. Those characteristics are fulfilled by many biologic meshes. Besides, biologic mesh can also promote cellular ingrowth, neovascularization, and potential regeneration of native tissue [6]. However, synthetic mesh remains in





Fig. 5. Post-operation 6 months, there was denser hair growth over the ALT flap.

widespread use despite the biologic mesh carries more advantages due to its exceptionally expensive cost. On top of that, patients' religious and moral convictions must be taken into account while deciding for a biologic mesh preoperatively. The index patient was a Muslim, thus, the porcine-sourced mesh must be avoided. We used an on-lay prolene mesh for our patient because the biologic mesh was not available in Malaysia. In the event of a breach in the peritoneal layer during wide local excision, we had planned to use a composite mesh instead to prevent adhesions of the intra-abdominal organs to the mesh. In order to ensure the on-lay mesh could resurface the entire abdominal wall, it was anchored from the left external oblique to the contralateral external oblique and the remaining right anterior rectus sheath. Quilting sutures were also used in multiple locations to decrease seroma formation besides from putting drains.

Regardless of the mesh type, the overlying soft tissue must deliver a stable cutaneous coverage and obliteration of dead space [4]. However, the patient developed marginal flap necrosis and surgical site infection with infected mesh. The patient subsequently underwent partial removal of the mesh and multiple wound debridement, rendering a defect over the cranial part of the wound. In order to achieve a satisfactory soft tissue closure, the right MICAP flap was recruited to close the defect over the epigastrium. SSG was harvested to cover the raw area over the right lower costal region. Post-operation 3 months, the patient noted denser hair over the ALT flap, this phenomenon has not been reported to date. Two postulations were made based on the clinical experiences. Firstly, It could be due to recurrent shaving. In addition, we hypothesized that the hormonal changes could be the culprit. However, the ALT flap was not shaved prior to or during the operation and the patient denied shaving it after discharge. The subsequent hormonal tests (serum TSH, cortisol, progesterone, estradiol and testosterone) were all within normal range. Further study is thus advised to elucidate the

underlying mechanism of the situation. Overall, the patient was still satisfied with his activities of daily living, self-image and subjective well-being. This case report has been reported in line with the SCARE 2020 criteria [7].

#### 4. Conclusion

Extra-skeletal Ewing Sarcoma is a rare clinical entity. A large anterior abdominal defect poses a challenge in soft tissue reconstruction. The goals of anterior abdominal wall reconstruction are restoration of integrity of the layers of the abdomen, prevention of visceral eventration, and provision of functional support. The use of combination of various technique, namely free ALT fasciocutaneous flap with AV loop graft, right MICAP flap and SSG in reconstruction ensures a satisfactory functional and aesthetic outcome.

#### Provenance and peer review

Not commissioned, externally peer reviewed.

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This work had no source of funding.

#### Ethical approval

No ethical approval required for the mentioned case report. However, permission was obtained from local administrative and this included consent from patient.

#### Consent

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 2013 and its later amendments. Informed consent was obtained from the patients family for being included in this case report prior to submission. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

Liu Y., Zosimo Ken L. Jimeno IV, Ramesh Sasidaran, Sentilnathan Subramaniam initiated and planned the case report. Liu Yi did the writing of the manuscript. Zosimo Ken L. Jimeno IV and Sentilnathan Subramaniam supervised, reviewed and edited the manuscript. Liu Yi were involved in the writing and providing the clinical data.

#### Registration of research studies

Not applicable.

#### Guarantor

Liu Yi will be the guarantor and accepts full responsibility for the work and/or the conduct of the study, had access to the data, and controlled the decision to publish at this given time of submission.

#### Declaration of competing interest

All authors declare that they have no conflict of interest.

#### Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1054/ams.2022.103171>

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