CASE REPORT

Metastatic Pleomorphic Dermal Sarcoma Involving Tonsil and Thyroid: An Unusual Presentation

Marek Solomianko[1], Irene Y Chen[2], Srinivas Kaza[3], Dongwei Zhang[4]

Abstract

Pleomorphic dermal sarcoma (PDS) is a rare dermal-based mesenchymal tumor that shares histologic characteristics with atypical fibroxanthoma (AFX), but demonstrates aggressive features increasing the risk of local recurrence or metastases. We report a unique case of a 62-year-old man with a recent history of left-groin PDS, now with a 2-cm pedunculated left tonsillar mass and two hypermetabolic nodules in the left thyroid. Microscopically, the tonsillar biopsy demonstrated infiltrating spindle cell neoplasm composed of highly pleomorphic tumor cells with abundant mitoses. The left hemithyroidectomy specimen also demonstrated similar pleomorphic spindled tumor cells infiltrating thyroid parenchyma with perineural invasion and focal osseous and chondroid components. Immunohistochemically, both tumors were diffusely positive for smooth muscle actin. Focal nonspecific positive staining with p53, SATB2, and S100 was observed in the thyroid tumor. Additional markers, including p40, CK5, p63, pancytokeratin, CAM5.2, S100, SOX10, Melan-A, HMB45, CD45, CD68, CD163, caldesmon, p16, TTF1, PAX8, and calcitonin, were all negative. INI-1 expression was retained. Molecular analysis showed that the tumor harbored *PDGFRA* deletion of exon 9 and amplification, *PTCH1* A300fs*24, *MDM2* amplification, *CDKN2A/B* loss, and *FRS2* amplification. Compared with the patient's groin tumor, the current tumors showed similar morphology. Overall, the histologic and immunohistochemical findings are consistent with metastatic PDS to the tonsil and thyroid. Although PDS is commonly found on the sun-exposed skin in the elderly, this case highlights an unusual primary site as well as peculiar locations for the metastasis of PDS. Treatment and management of the patient are also considered.

Keywords: Atypical fibroxanthoma, Metastasis, Pleomorphic dermal sarcoma, Thyroid, Tonsil.

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Background

Pleomorphic dermal sarcoma is a rare dermal-based mesenchymal neoplasm that was once under different nomenclature causing confusion in regard to identification and management of such tumors. Pleomorphic dermal sarcoma was first proposed by Dr CD Fletcher describing tumors that share histologic characteristics with AFX, but demonstrate aggressive features, such as deep subcutaneous involvement, necrosis, perineural invasion, or lymphovascular invasion, increasing the risk of local recurrence or metastases. Pleomorphic dermal sarcoma and AFX have similar clinical presentations, both of which are commonly found as rapidly growing masses with ulceration on sun-damaged skin in the head and neck region with a strong male predilection and age of onset at 80 years.[1,2] Differentiating PDS from its less-aggressive counterpart is vital since PDS has higher metastatic potential and commonly metastasizes to lung, bone, liver, other cutaneous locations, and lymph nodes.[3,4] Herein, we describe a case of PDS arising from the inguinal region with widespread metastases in an unusual distribution.

Case Description

A 62-year-old male with no significant past medical history presented with a tender inguinal mass for the past several months, which has failed medical management. Computed tomography (CT) scan confirmed a 1.2-cm dermal/superficial subcutaneous enhancing nodule of the distal anterior right-abdominal wall without any lymphadenopathy or lung nodules in the lower lung fields (Figs 1A and B). The patient was referred to have the mass completely excised and sent to pathology for histologic analysis (Fig. 2). Hematoxylin and eosin (H&E)-stained sections of the groin mass revealed a dermal-based lesion consisting of pleomorphic spindle cells with necrosis and numerous mitoses. The overlying epithelium was intact and uninvolved. Immunohistochemically, the spindle cells are diffusely positive for smooth muscle actin (SMA) while negative for AE1/AE3, S-100, DOG-1, p40, CK5, p63, keratin cocktail, SOX10, Melan-A, HMB45, CD45, CD68, CD163, ERG, CD31, and caldesmon. INI-1 expression was retained. The overall findings are consistent with PDS. No adjuvant therapy was recommended at this point due to the negative margins achieved with the resection and the small size of the tumor. At 4 months postop, no evidence of metastatic pulmonary disease was identified on the

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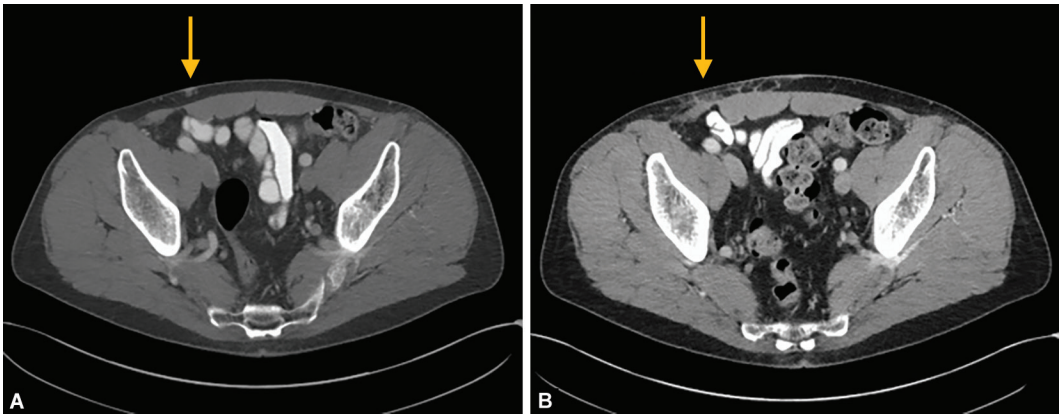
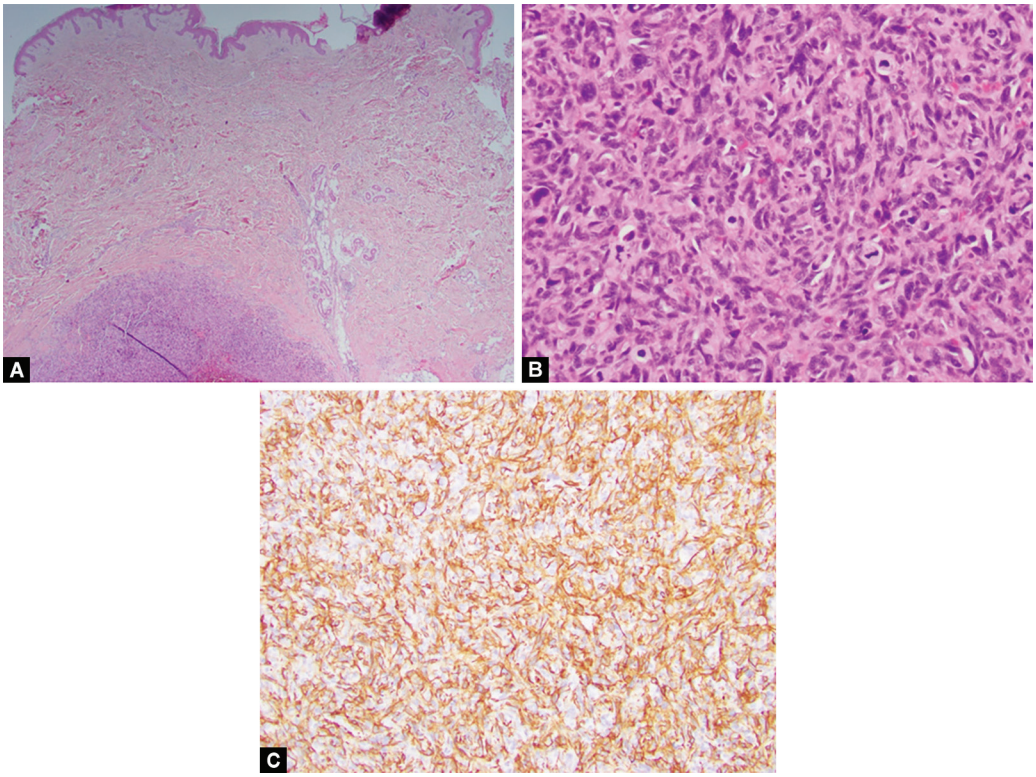


Figs 1A and B: CT abdomen and pelvis with contrast, horizontal view. (A) 1.2 cm dermal/superficial subcutaneous enhancing nodule of the right, distal anterior abdominal wall; (B) Postoperative inflammatory changes and stranding in the area where the resection was performed



Figs 2A to C: Histology and immunoprofile. (A) The tumor is in the dermis with no epidermal involvement (H&E; 20×); (B) The tumor cells are spindled, irregularly dispersed, and admixed with abundant mitoses (H&E; 200×); (C) Immunohistochemistry shows tumor cells in the groin are positive for SMA (100×)

surveillance chest radiograph. A surveillance abdominal CT revealed postoperative changes and stranding in the surgical area (Fig. 1B).

At 8 months postop, the patient presented to the otolaryngologist with worsening dysphagia and globus sensation. A 2-cm pedunculated mass emanating from the left base of the tongue/tonsillar fossa was identified. The excisional biopsy demonstrated infiltrating spindle cell neoplasm composed of highly pleomorphic tumor cells with abundant atypical mitoses (Fig. 3). The histologic findings and immunoprofile of the specimen were consistent with metastases from the patient's known history of PDS.

Evidently, the patient had been hospitalized for acute pancreatitis just prior to his presentation to the ENT. An abdominal CT scan was performed during his workup that showed a lesion in the pancreas. A biopsy of the lesion was consistent with similar high-grade spindle cell sarcoma. A subsequent PET scan showed hypermetabolic foci in the pancreas, thyroid, and sacrum (Figs 4A to C). The patient was then started on systemic chemotherapy as well as targeted radiotherapy.

After two rounds of chemotherapy, the patient again presented to our clinic with a new complaint of pain at the anterior left-sided neck. Thyroid ultrasound showed a 2.5-cm left-sided thyroid nodule with irregular borders. Correlation with previous CT scans and PET scans confirmed the presence of a hypermetabolic nodule in the same area (Fig. 4A). After discussions

with oncology, it was collectively agreed to monitor the thyroid lesion closely with serial ultrasound exams and continue with the chemotherapy regimen.

After two-and-a-half months with minimal change in the thyroid nodule, the patient presented with new-onset hoarseness. Repeat ultrasound showed that the lesion had grown to 2.9 cm and flexible fiberoptic laryngoscopy diagnosed a new left vocal fold paralysis. The patient consented to a left hemithyroidectomy. Intraoperatively, the left recurrent laryngeal nerve was encased in the tumor and could not be salvaged. Tumor invasion extended to the infrahyoid muscles and the cricothyroid joint. Several central compartments and paratracheal lymph nodes were resected. The tracheal wall remained intact. The left hemithyroidectomy specimen showed similar highly pleomorphic spindled tumor cells infiltrating thyroid parenchyma with focal osseous and chondroid components and abundant perineural invasion (Fig. 5). A postoperative PET scan showed no further FDG enhancement in the area of the thyroid (Fig. 6).

Discussion

Pleomorphic dermal sarcoma is a rare neoplasm that most frequently presents in the extremities or head and neck, and most often in people between the ages of 50 and 70-years-old.[5,6] Histologically, it consists of spindle-shaped cells containing pleomorphic epithelioid and giant multinucleated cells in varying proportions, along with numerous mitotic figures and varying levels of necrosis.[4] In the past, it has been given many different names including cutaneous undifferentiated pleomorphic sarcoma and malignant fibrous histiocytoma. Pleomorphic dermal sarcoma has a metastatic risk estimated to be less than 5%, however, due to its rarity and lack of homogeneous criteria used for classification, the true rate is not exactly known. According to a retrospective study of 18 cases performed by Tardio et al., the metastatic potential may be higher than previously thought.[4] Another report by Jeong et al. reported a local recurrence rate of 19–31% and a metastatic rate of 31–35% even after tumor resection with clear margins.[3] The most common sites of metastasis are lung (90%), bone (8%), liver (1%), skin, and lymph nodes.[1,3]

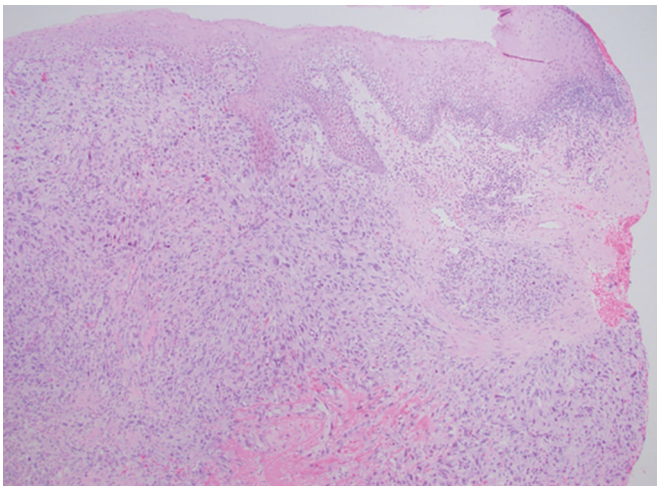
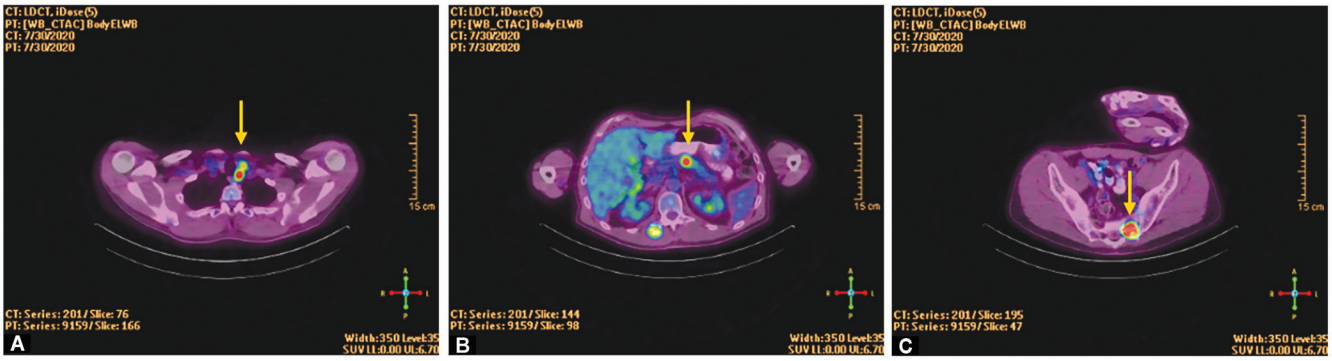


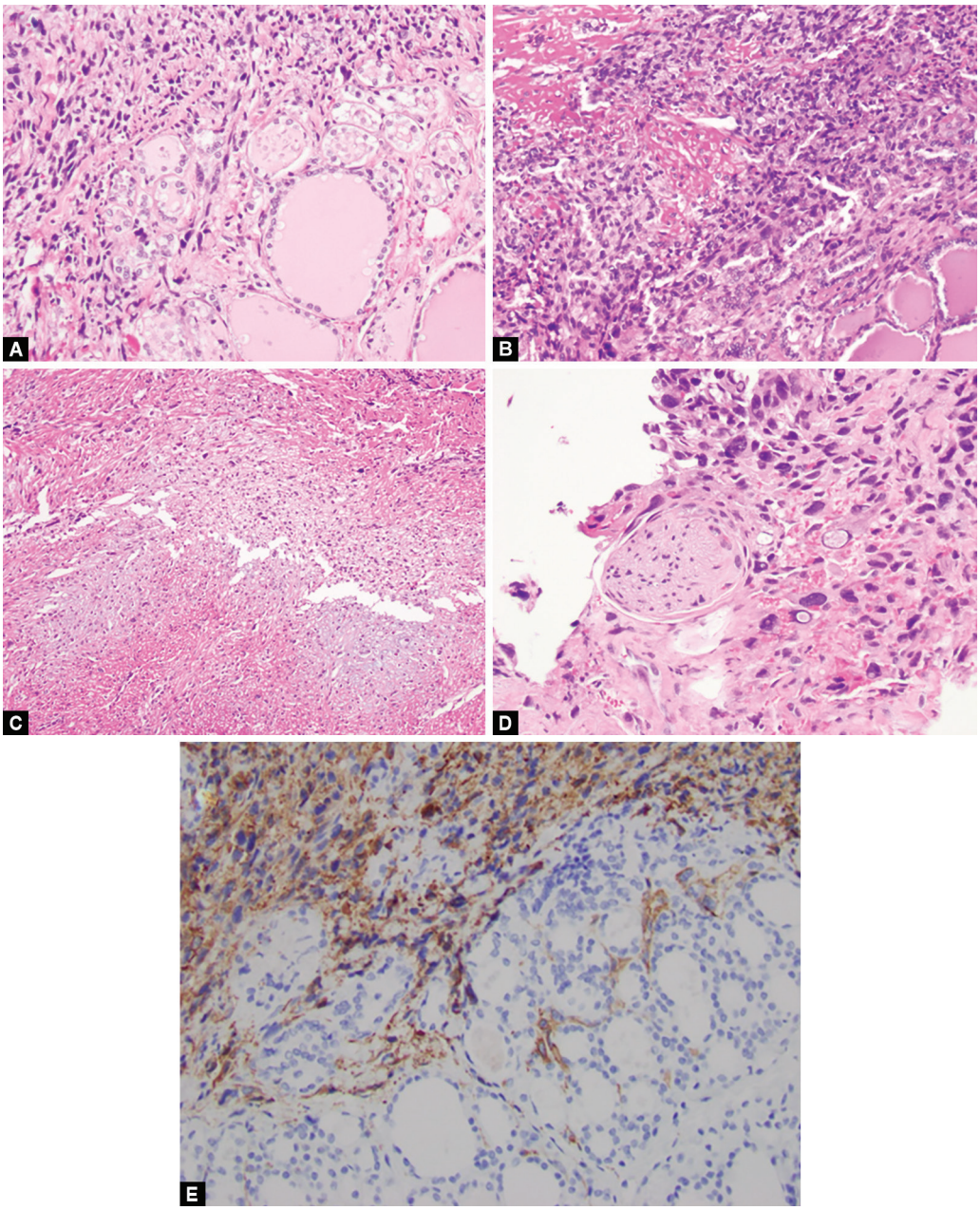
Fig. 3: The tumor cells invade into tonsil (magnification: 40×)

In this presented case report, a pertinent question is whether the patient would have benefitted from adjuvant radiotherapy or chemotherapy after the initial inguinal surgery. Some indications for postoperative radiotherapy include unresectable primary tumor, locally recurrent, or regionally metastatic disease,[7] none of which were apparent factors in this case. Another study recommends that for soft-tissue sarcomas of the head and neck, postoperative adjuvant radiotherapy is indicated for patients with low-grade tumors with close (<1 cm) or positive margins, as well as those with high-grade tumors.[8] Complete compartmental resection of tumors in the head and neck is usually not feasible, and even with wide-margin resection the recurrence rates may be as high as 50%, and therefore, most patients should receive adjuvant radiotherapy.[8] The role of adjuvant chemotherapy is unclear as well, however, there are several reports that show some promise. A meta-analysis of sarcomas of the extremities showed that doxorubicin-based adjuvant chemotherapy improved both local control and distant metastases-free survival.[8] Other studies show adjuvant chemotherapy being used in cases of multiple metastases, consistent with the patient's current therapy.[3]

Here, we present a series of PET/CT images that show how the patient's cancer responded at various foci (thyroid, pancreas, and sacrum) to the combination of chemotherapy and targeted radiotherapy (Figs 7A to C, Figs 8A to C, Figs 9A to C). The first image in each group shows scans prior to the patient receiving any adjuvant chemoradiotherapy (Figs 7A, 8A, 9A). The second and third images in each group show scan after adjuvant treatment was started and are taken 4 months apart (Figs 7B and C, Figs 8A to C, Figs 9B and C). There was a partial response in the thyroid foci to systemic chemotherapy visible when comparing Figures 7A and B. There was complete resolution of the thyroid foci after left hemithyroidectomy visible when comparing Figures 7B and C.



Figs 4A to C: PET/CT (FDG). (A) Horizontal view showing 2 hypermetabolic foci in the thyroid; (B) Horizontal view showing hypermetabolic focus in the pancreas; (C) Horizontal view showing hypermetabolic focus in the sacrum



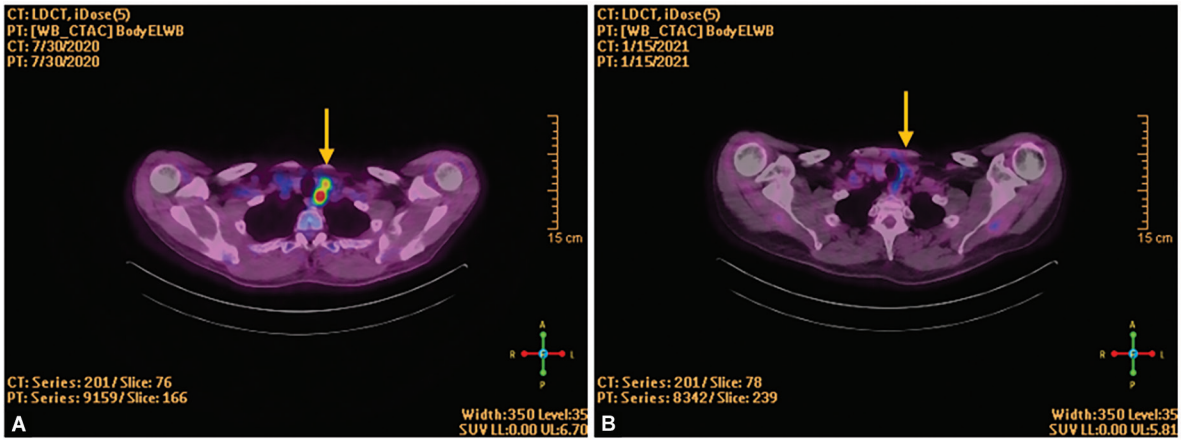
Figs 5A to E: Histology of left hemithyroidectomy. (A) The tumor cells infiltrate thyroid parenchyma (200×); (B) Focal osseous component (200×); (C) Focal chondroid component (100×); (D) Representative picture of perineural invasion (200×); (E) Immunohistochemistry shows tumor cells in the thyroid are positive for SMA (200×)

The pancreatic lesion showed a complete response to treatment (Figs 8A to C). The sacral lesion showed a partial response to treatment (Figs 9A to C).

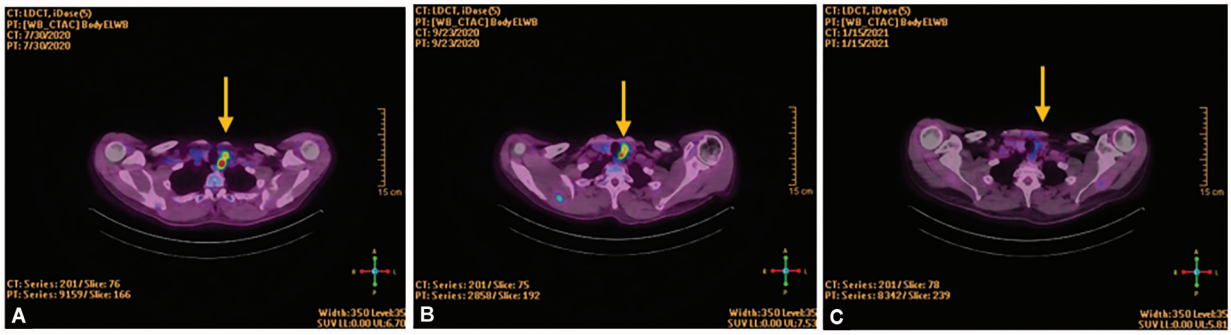
The atypical pattern of metastatic spread is the most salient feature of the case. We were unable to find previous reports that showed metastases to the posterior oropharynx or to thyroid tissue. As stated previously, the most common sites of metastasis for PDS include the lung (90%), bone (8%), liver (1%), skin, and lymph nodes.[3,5] In the case presented, the presumed primary site was a cutaneous lesion in the inguinal region. From there, multiple subsequent lesions were discovered at the base of the tongue/tonsillar fossa, pancreas, esophagus, paraspinal musculature, sacrum, pubic rami, psoas muscle, and ultimately thyroid gland.

Conclusion

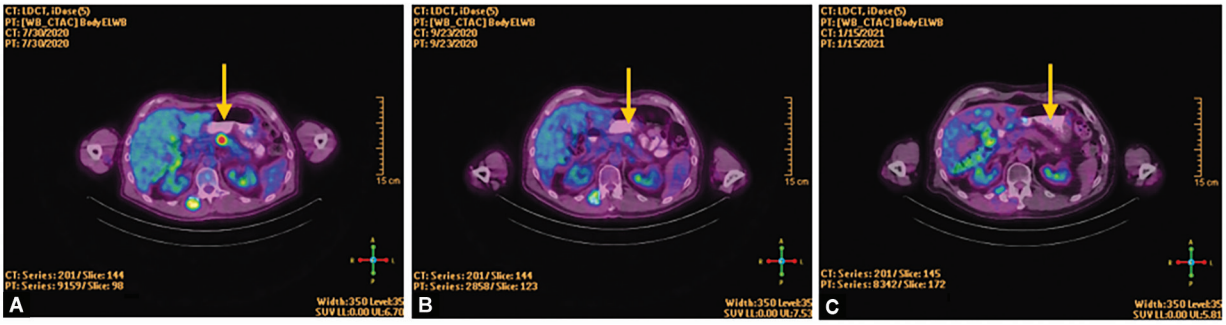
Pleomorphic dermal sarcoma, also known as cutaneous undifferentiated pleomorphic sarcoma, is a rare tumor of mesenchymal origin. We present the case of a 62-year-old male with a PDS arising from cutaneous tissue in the inguinal region which ultimately metastasized to the base of the tongue/tonsillar fossa, pancreas, esophagus, paraspinal musculature, sacrum, pubic rami, psoas muscle, and thyroid gland. In this challenging case, the patient did not receive postoperative adjuvant radiation



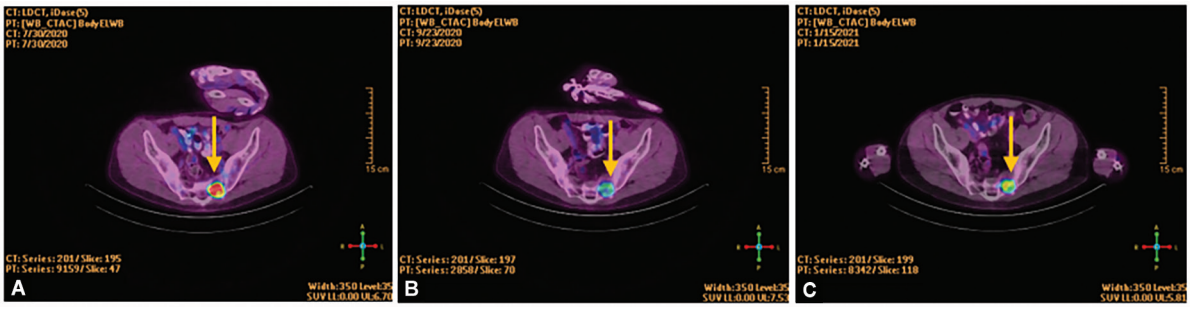
Figs 6A and B: PET/CT (FDG). (A) 2 hypermetabolic foci in the thyroid prior to hemithyroidectomy; (B) Postoperative hemithyroidectomy image showing complete removal of the 2 prior hypermetabolic foci



Figs 7A to C: PET/CT (FDG) images showing the response of the thyroid lesions to chemoradiotherapy and hemithyroidectomy. (A) Prior to any chemoradiotherapy; (B) After chemoradiotherapy; (C) After left hemithyroidectomy



Figs 8A to C: PET/CT (FDG) images showing the response of the pancreatic lesion to chemoradiotherapy. (A) Prior to any chemoradiotherapy; (B) After chemoradiotherapy; (C) 4-month follow-up scan



Figs 9A to C: PET/CT (FDG) images showing the response of the sacral lesion to chemoradiotherapy. (A) Prior to any chemoradiotherapy; (B) After chemoradiotherapy; (C) 4-month follow-up scan

or chemotherapy initially, but ultimately became a candidate for both adjuvant modalities once metastatic disease was identified.

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