Case of the Week

COMBINING SURGERY AND BRACHYTHERAPY FOR AN UNUSUAL METASTATIC SITE OF LEIOMYOSARCOMA

Leiomyosarcomas are tumours of the smooth muscle cells that may originate in any location, but most often arise in the uterus, gastrointestinal tract and soft tissue. Primary infundibulopelvic ligament leiomyosarcomas (LMS) are extremely rare and constitute about 1% of the gynaecological malignancies (2). Common sites of metastasis are lung, liver, pelvic lymph nodes, and vertebral and long bones, though atypical patterns of recurrence have been described. Metastasis of these tumours to the neck is uncommon, even so in absence of lung or liver metastasis (1).

We report a case of unusual presentation, in a premenopausal 27 years' young lady, presented in January 2019 with feeling of heaviness in left flank. She underwent laparoscopy for suspected torsion of ovary. Incidentally, a soft tissue mass near left lateral pelvic wall was found and biopsied. The histopathology revealed leiomyosarcoma. She underwent left oophorectomy with pelvic soft tissue mass excision and left pelvic lymph node dissection in March 2019. Histopathology was pleomorphic spindle cell sarcoma arising from left infundibulopelvic ligament with deposits over the left ovary. She received adjuvant chemotherapy with Gemcitabine and Docetaxel till July 2019. In June 2020, the follow up CT-scan showed soft tissue lesion adherent to the uterus for which she underwent total abdominal hysterectomy, excision of pararectal deposit and excision of adherent omentum. The histopathology showed a 4.7 cm leiomyosarcoma involving uterus with pararectal deposit. She received adjuvant radiation therapy to pelvis, however during radiation therapy, she noticed small swelling in left neck. The ultrasonography of neck showed a 2.9 x 2.3 x 1.3 cm level-II lymph node suggestive of metastatic deposit. Following biopsy confirmation, she was given single agent Adriamycin, but the size of the neck node increased while on chemotherapy. PET CT scan done in January 2021 showed 3 x 3 cm partially cystic FDG avid (SUV 11.03) left cervical level II node, partially effacing the internal jugular vein, without any evidence of disease elsewhere. We planned her for selective neck dissection (level I-III) and intraoperative brachytherapy catheters placement.

At surgery, we observed the nodal masses to be firm to hard in nature. It involved a few medial fibres of the sternocleidomastoid muscle and partially engulfed the spinal accessory nerve. The nerve was dissected off the tumour without leaving any gross tumour tissue. Complete dissection of level I-III was done. The final histopathology revealed leiomyosarcoma, expressing SMA and Desmin and immunonegative for CD 34, CD 117 and S100. Ki-67 proliferative index was 20-25%.
She received 35 Gy in ten fractions (3.5 Gy per fraction twice a day 6 hours apart) high dose rate (HDR) brachytherapy with Iridium 192 source, starting after 3rd day of surgery. She tolerated the treatment well without any undue reactions.

Combining surgery and intraoperative brachytherapy helped her to complete her treatment early, reducing her hospital stay. It is important to be aware of atypical patterns of recurrence. Although metastatic LMS is associated with poor prognosis, the solitary metastasis possibly shows better clinical outcomes. The present case showed solitary left cervical level II nodal metastasis without other metastatic foci. Given the rarity of metastasis to this region, there is a lack of evidence supporting a specific treatment. Management is therefore personalized based on clinical condition, site, and size of metastasis. Optimal treatment strategies should be adopted for patients with good performance status and who are suitable for surgery, chemotherapy and radiotherapy.

Points:

- Primary head and neck leiomyosarcomas are rare since smooth muscle is sparse in head and neck and found mainly in the tunica media of blood vessels, erector pili musculature of skin, myoepithelial cells of mucous glands and pluripotent mesenchymal cells (4). Hence, a neck mass with the diagnosis of leiomyosarcoma should raise a strong suspicion of disease primary elsewhere and needs appropriate imaging.

- Neck metastasis of leiomyosarcoma has been reported to have good response to treatment by surgery and brachytherapy (2).

- Primary leiomyosarcoma of the pelvic organs can rarely metastasize to the neck, without having lung or liver metastasis. The possible route of metastasis from non-head and neck sites has been postulated to be Batson's vertebral venous plexus, connecting the cervical, thoracic and lumbar region (2, 3).

- Head and neck leiomyosarcomas are of high grade and occur in the skin and soft tissues as the primary site (1). It is important to distinguish a primary subcutaneous leiomyosarcoma from a secondary leiomyosarcoma because treatment and prognosis are very different (4).

References:

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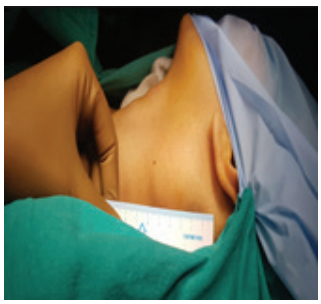

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