

Malignant triton tumor of the neck: A case report

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Abstract: Malignant Triton tumor (MTT) is a rare variant of Malignant Peripheral Nerve Sheath Tumors (MPNST) with striated muscle formation. The diagnosis of MTT must meet the diagnostic criteria of MPNST and proven rhabdomyoblast differentiation. This case is discussed because of its rare incidence, which is 5-10% of all MPNSTs. The prognosis in most MTT patients is poor due to its rarity. Malignant Triton tumor (MTT) is a rare variant of malignant peripheral nerve sheath tumor. A 34-year-old woman came to the Head and Neck Surgery clinic of RSUD Dr. Soetomo Surabaya with complaints of a lump on the left upper front neck. This painless lump was small, marble-sized, skin-colored, and had enlarged rapidly over the last year. On physical examination, a skin-colored mass was found in the anterior colli region sinistra, which extended to the infraauricula sinistra, measuring approximately 20 x 20 x 15 cm with a hard solid consistency, partially spongy, flat surface, and indistinct borders. The patient underwent wide excision, vascular exploration, and continued with radiotherapy. The prognosis in most MTT patients is poor, and due to its rarity, studies with a large number of cases are lacking. MTT is a rare variant of MPNST with rhabdomyoblastic differentiation that behaves more aggressively than typical MPNST, thus having a worse prognosis. Treatment modalities, especially in advanced and metastatic cases, still lack standardized management or guidelines for the management of MTT. Until now, surgical excision and adjuvant radiotherapy have been the main therapeutic options for MTT.

Keywords: *Head and Neck Malignancy, Malignant Triton Tumor, Nerve Sheath Tumor.*

1. Introduction

Malignant Triton Tumor (MTT) is a rare variant of Malignant Peripheral Nerve Sheath Tumors (MPNST) with striated muscle formation. MPNSTs stem from the neuroectoderm and arise from Schwann cells of peripheral nerves or nearby cells, either through independent perineural differentiation or within an existing neurofibroma. MTTs are named after the triton salamander that has the ability to develop supernumerary limbs composed of a mixture of neurogenic and muscular components after implantation of sympathetic nerves on its dorsal surface [1, 2].

MTT comprises 5-10% of MPNST cases and typically occurs between the third and fifth decades of life. Studies indicate that 50- 70% of these tumors develop in patients with neurofibromatosis type 1 (NF-1). MTT exhibits highly aggressive behavior, with a local recurrence rate of 43-50%. The reported five-year survival rate is approximately 14%, significantly lower than the 34-60% observed in MPNST cases overall [2-5].

MTT presents significant diagnostic challenges for pathologists due to its histological complexity, often leading to misdiagnosis as conventional MPNST. The diagnosis of MTT must fulfill the diagnostic criteria of MPNST and proven rhabdomyoblast differentiation. Diagnosis is established through morphological evaluation, supported by immunohistochemical findings, including positive staining for myogenin, S-100 protein, and desmin [4, 5].

While MTTs can develop sporadically, about one-third of cases occur in the head and neck region. The estimated clinical incidence of MTT is fewer than 1 per 1,000,000 people. Current management typically involves wide excision or tumor debulking with negative margins, followed by radiotherapy. While chemotherapy has been explored, its efficacy remains unproven. Given the limited number of reported cases and the absence of large cohort studies, further research is needed to better understand the clinical features of MTT. In this context, the authors present a case of MTT treatment at RSDS and provide a literature analysis [6, 7].

2. Case Report

A 34-year-old woman came to the Head- Neck Surgery outpatient clinic of RSUD Dr. Soetomo Surabaya with a chief complaint of a lump on the left upper neck since 15 years ago. The lump was initially small, marble-sized, skin-colored, movable and painless. About 1 year before the patient sought treatment because the lump was getting bigger up to the size of a fist and felt painful. The patient then went to Genteng Hospital and FNAB was performed. Because the results of the cytological examination were not representative, 2 weeks later an open biopsy was performed and showed a fibroadenoma. Since the biopsy was performed, the lump was felt to be getting bigger and within 6 months, making the patient hard to turn his head to the left. The patient also complained that he could not make a loud voice. There were no complaints of swallowing disorders, bone pain, shortness of breath, coughing, headache, nausea, vomiting, and abdominal pain. On physical examination, there was a mass in the anterior colli region sinistra extending to the infraauricula sinistra sized +/- 20 x 20 x 15 cm, hard consistency, indistinct boundaries, and no neck lymphnode enlargement palpable. The patient was clinically diagnosed with a soft tissue malignant tumor.



Figure 1.
The mass on left neck region.

Head and Neck CT Scan with contrast showed a solid mass (38 HU) with a necrotic component (23 HU) inside, well-defined borders, irregular edges, size +/- 18.2x16.6x17.6 cm in the soft tissue of the colli region sinistra which on contrast enhancement (76 HU), with the infero-posterior side pressing the prevertebral space, masticator space, constricting v. jugularis interna and a. carotid

communis to the medial side, the anterior side pressing the submandibular gland attaching the extrinsic muscles to the medial side, left internal jugular and carotid communis artery to the medial side, the anterior side pressing the submandibular gland, attaching the extrinsic m. (m. genioglossus, m. sternohioid) with a firm border, medial side pressing and constricting the laryngeal inlet with a narrowed lumen of ± 0.85 cm and pressing the left lobe of the thyroid with a firm border. Multiple lymph nodes were seen in the peritumoral area and in the right upper jugular. There was also a solid mass (30 HU), firm borders, regular edges, in the right side mediastinum medius, size 3.7 x 2.6 x 4.7 cm, it attached to the aortic arch and inferior vena cava with firm borders. From contrast CT scan of the thorax, a solid mass (33 HU) with firm borders, regular edges in the right mediastinum medius measuring 2.75x3.55x5.61cm no contrast enhancement, and the mass was attached to the SVC, right brachiocephalic trunk and trachea on the right side. There was also a semisolid mass (23HU) in the paravertebral, inside and widening the foraminal canal and partially in the intradural-extramedulla as high as VTh 12- VL1 on the right side with a size of 4.3x5.2cm which on contrast administration showed slight contrast enhancement.

The biopsy specimen from the referring hospital revealed a spindle mesenchymal tumor, with a differential diagnosis including a solitary fibrous tumor or low- grade myxofibroma.

Immunohistochemical analysis of the same sample confirmed the presence of Vimentin positivity at the cytoplasm of tumor cells and CD34 positivity at the tumor cell membrane. Additionally, EMA, SMA, and desmin were negative in the cytoplasm of tumor cells, while myogenin was negative in the nucleus. Based on these findings, the anatomical pathology diagnosis was determined to be a solitary fibrous tumor.

From the clinical, radiological and pathological findings, the patient was diagnosed as neurofibromatosis (neck, mediastinum, paravertebra Th12-L1) with malignant differentiation in the neck lump (T4bN0M0). In this patient, surgery was performed with excision of the tumor in the colli regio first with consideration of the mass possibly having malignant differentiation. Intraoperatively, a hard solid mass, well-defined, fixed to the base, with a size of 20x25cm, with a. carotid

communis, a. carotid interna, a. carotid externa, v. jugular interna and v. jugular externa were found within the tumor and there was a rupture at the bifurcation of a. carotid communis. Tumor excision was performed with preservation of a. carotis communis. However, because the distal part of a. carotis externa sinistra was not found during identification, ligation of a. carotis externa sinistra was performed. The surgical wound was then closed with primary sutures and a #12fr redon drain was placed.

The tissue obtained after the operative procedure was a tumor weighing 2,879 g, with a size of 25x25x21 cm, which in slices appeared to be a white-gray mass with necrotic, capsular, partially intact parts. The slice surface of the tumor was palpably slippery coated with a mucin-like liquid.

Microscopic examination shows pieces of tissue partially coated with skin, with well- defined tumor growth arranged in a light- dark pattern (“marble-like”), the fascicles are partially arranged palisading. The tumor consists of oval to spindle nucleated cells, pleomorphic, coarse chromatin, narrow cytoplasm. Blood vessels with hemangiopericyte-like appearance. Stroma in the form of partially hyalinized myxoids, rhabdomyosarcomatous heterologous differentiation consisting of cell proliferation of oval-spindle nucleated cells, coarse chromatin, moderate to broad cytoplasm, eosinophilic, and osteosarcomatous differentiation consisting of a “lace-like” osteoid matrix. Areas of necrosis are also visible. Mitosis 20/10 HPF. There was no lymphangioid invasion or perineural invasion. Distance from the tumor to the nearest resection edge: coincident. There was no metastasis in the 5 KGB nodules found. And from immunohistochemical examination, Vimentin was positive in the cytoplasm of tumor cells, S100 was positive in the cytoplasm of tumor cells and Desmin was positive in the cytoplasm of tumor cells, which corresponded to the picture of Malignant Triton Tumor.

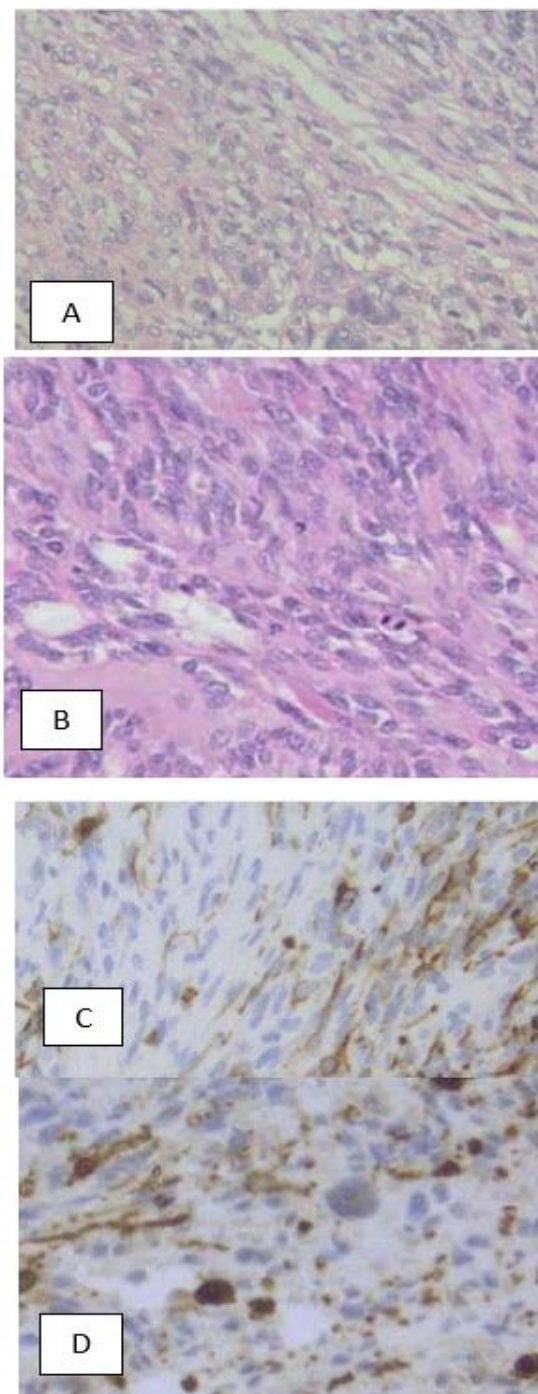


Figure 2.
Tumor under the microscope.

At 3 weeks postoperatively, an evaluation was carried out on the patient and was no infection in the surgical wound, there was no mass in the surgical wound or colli region and there were no signs of impaired vascularization or nerve disorders in the head and neck area.

The next patient was planned for radiotherapy to the left neck area with a radiation dose of 50-70

Gy in consideration of the high local recurrence of MTT. For T12-L1 mediastinal and paravertebral mass in the following patient, it was planned to confirm the diagnosis first. Six months post-surgery, before receiving adjuvant radiotherapy, the tumor in the neck arose and enlarged again with the size as before surgery, and part of the tumor penetrated the skin. Due to impending airway obstruction, the patient underwent tracheostomy while urgently requesting radiotherapy. A few days after tracheostomy, the patient passed away due to advanced MTT. A & B The appearance of Malignant Triton Tumor histologically; C. S100 positive in tumour cell; D. Desmin positive in tumor cytoplasm

3. Discussion

Malignant Triton Tumor (MTT) is an uncommon form of malignant peripheral nerve sheath tumor (MPNST) that exhibits rhabdomyoblastic differentiation. The name "triton" is derived from the triton salamander, which can regenerate additional limbs containing both neural and muscular components when sympathetic nerves are introduced to its dorsal region. This tumor was initially recognized as a disease in 1932 and later classified as Malignant Triton Tumor in 1973 [2, 3, 8]. MTT accounts for <10% of MPNST. NF-1 (von Recklinghausen's disease) has been associated with an increased risk of MPNST and MTTs. MTTs can appear in patients with NF-1 as well as sporadically. Two-thirds of MTT cases have been reported to be associated with NF-1. MTTs with NF-1 are seen mainly in males of the younger age group (28-36 years); whereas sporadic forms are seen in females of the older age group (40-44). When presenting in sporadic form, other spindle cell sarcomas such as fibrosarcoma, malignant fibrous histiocytoma, and rhabdomyosarcoma can be a differential diagnosis. The tumor develops after a long latent period of 10-20 years. In our case, the tumor appeared sporadically when the patient was in her 2nd decade of life and developed after 15 years [4, 8, 9].

Woodruff, et al. [10] proposed three criteria for the diagnosis of this disease: 1) arising along peripheral nerves ganglioneuromas or in patients with neurofibromatosis type 1 (NF-1); 2) having the growth characteristics of Schwann cell tumors; and 3) the presence of rhabdomyoblasts within the tumor. Daimaru, et al. [11] later expanded the definition to include sporadically occurring cases in patients without NF-1, according to which, malignant Schwannoma with focal rhabdomyoblasts is microscopically comparable to tumors composed predominantly of rhabdomyoblastic differentiation with focal Schwann cell elements occurring within nerve cells or in patients with NF-1. Regarding the histopathogenesis of MTT, the most plausible explanation is the fact that multipotent neural crest cells from the ectomesenchyme can differentiate into neuronal and muscle components in distinct ways. Additionally, it has been proposed that motor neurons can induce Schwann cells in neurogenic tumors to develop into rhabdomyoblastic components. Kamperis, et al. [12] showed in a mouse model that neoplastic Schwann cells have the ability of mesenchymal differentiation into rhabdomyoblasts. Recent research on cytogenetics has revealed several karyotypic changes associated with this tumor. There is a breakpoint in the 11p15 gene that is thought to be the region where myogenic differentiation occurs. This gene is thought to be responsible for rhabdomyoblastic differentiation. While amplification of the c-myc oncogene is thought to be responsible for its aggressive biological behavior. In most reported cases, MTTs are located in peripheral nerves, usually close to the spine in the head and neck region. Some cases were found to arise in the mediastinum and extremities. Areas that have also been reported to harbor MTTs but rarely are the buttocks and retroperitoneal region. In this case, a mass was found in the head and neck region as well as the mediastinum and paravertebral Th12-L1. For mediastinal tumors, it is still limited to radiological examination (CT scan of the thorax with contrast), so histopathological examination is still needed to rule out other differential diagnoses of mediastinal tumors. Likewise, the mass at paravertebra Th12-L1 still requires further evaluation [2, 4, 5, 7, 12]. The diagnosis of MTT is based on pathologic examination. The histologic presentation on which the diagnosis of MTT is based is the proliferation of atypical spindle cells present within the abundant myxoid stroma. The bigeminal appearance of tumor origin is the basis for accurate diagnosis. Currently, the diagnosis of MTT is usually made after tumor resection with histopathology supported

by positive S-100 protein immunohistochemistry. The morphologic features are intermittent hypocellular and hypercellular areas, the appearance of a comma/bullet-shaped nucleus that is thin and wavy in the hypocellular area, the presence of palisade-arranged nuclei, prominent thick-walled blood vessels, and the presence of rhabdomyoblasts. Tumors with these features show positive S-100 protein in 50- 90% of cases. Rhabdomyoblasts are positive for immunohistochemical stains such as desmin, myogenin, and myo-D1. Interestingly, in this case, needle biopsy and incisional biopsy of the tumor showed a solitary fibrous tumor. This result was also confirmed by immunohistochemical examination of the same sample. However, from the pathological examination of the surgical specimen, differences were found where there was a picture of bright dark areas (“marble-like”), the fascicles were partially arranged palisading and in the stroma there was rhabdomyosarcomatous differentiation and osteosarcomatous differentiation. And from immunohistochemical examination, S100 and Desmin were positive in the cytoplasm of tumor cells which supported the MTT diagnosis. This may be because the tumor was initially a solitary fibrous tumor suspected of being an NF-1 which then differentiated into an MTT. However, due to the large size of the tumor, sampling for needle biopsy or incisional biopsy may not represent the entire tumor because it is possible that the part taken is the part that has not undergone differentiation [4, 13]. As MTT cases are still very rare, treatment modalities, especially in advanced and metastatic cases, there is still no standardized management or guidelines for management in these cases. As with most soft tissue sarcomas, the most effective therapeutic strategy for MTT is complete tumor resection. The microscopic status of the resection margin has a significant impact on local recurrence and survival. Enneking, et al. [14] described four types of histologic margins: intralesional, marginal, extensive and radical. Radical excision, radical excision followed by irradiation and chemotherapy, and excision with high-dose radiation are among the therapeutic recommendations provided by numerous research. However, the recommended course of treatment is tumor excision with broad margins followed by radiotherapy, much like in other sarcoma instances [1, 2, 14].

The role of chemotherapy methods in MTT instances is unclear. According to some recent publications, micrometastasis can be overcome with adjuvant chemotherapy and neoadjuvant therapy. More thorough second-stage surgical excision should always be carried out in cases with poor cytoreduction. The first-line chemotherapy in the suggested regimens is PEI (cisplatin, etoposide, and ifosfamide), whereas the second-line treatments are either IA (ifosfamide and adriamycin) or MAID (mesna, doxorubicin, ifosfamide, and dacarbazine). Another case report described an MTT patient who reacted well to treatment with interferon- α as well as interferon- γ as well as isotretinoin (retinoid analog). In this patient, tumor excision was performed on the basis of the initial diagnosis of a solitary fibrous tumor and because of the size of the tumor and the difficulty in determining the margins during surgery. On pathologic examination of the surgical specimen, the edge of the mass was found to coincide with the edge and base of the resection. This is our consideration for immediate radiotherapy in the postoperative area to avoid local recurrence. For management of mediastinal and paravertebral Th12-L1 tumors, we are still waiting for further diagnostics for pathological examination and evaluation of tumor resectability with consideration that currently the mediastinal paravertebral Th12-L1 tumor does not cause symptoms in the patient. The patient will be considered for radiotherapy of the mediastinal tumor if superior vena cava syndrome is found [4, 9, 12, 15]. The prognosis for MPNST is generally known to be poor, with a mortality rate within the first 2 years after diagnosis of 39-63%. MTT behaves more aggressively than MPNST in general, with 2-year and 5-year survival rates of 15% and 11% respectively. From a study conducted by McConnell and Giacomantonio on 24 MTT patients, the median survival time of patients with MTT was 13 months, the overall local recurrence rate was 50% and the median time to progression was 6 months. This study suggested surgical resection and adjuvant radiotherapy as the gold standard for the management of MTT. The study also concluded that conventional chemotherapy does not provide much benefit in MTT patients. The fact that MTT advances quickly in its early stages and is more likely to experience local recurrence and blood-borne

metastases are some factors that could account for the variation in prognosis between MTT and MPNST in general. MTT is more prevalent in older individuals, more frequently occurs in the head, neck, and body region, and results in larger tumors than MPNST. Furthermore, the metastasis-free time is shorter in MTT patients [3, 5, 16].

4. Conclusion

Malignant Triton Tumor (MTT) is a rare variant of malignant peripheral nerve sheath tumors (MPNSTs) with rhabdomyoblastic differentiation that behaves more aggressively than typical MPNSTs, thus having a worse prognosis. The diagnosis of MTT is based on pathological examination. Due to its rarity, treatment modalities, especially in advanced and metastatic cases, there is still no standardized management or guidelines for the management of MTT. Until now, surgical resection and adjuvant radiotherapy have been the main therapeutic options for MTT.

Transparency:

The authors confirm that the manuscript is an honest, accurate, and transparent account of the study; that no vital features of the study have been omitted; and that any discrepancies from the study as planned have been explained. This study followed all ethical practices during writing.

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