

Leiomyosarcoma of the parotid gland metastatic to the scalp: A rare primary location with unusual metastatic lesion

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ABSTRACT

We are reporting an extremely rare case of leiomyosarcoma of the parotid gland in a 33-year-old-woman, who was initially treated with surgery and post-operative radiotherapy. The patient developed an isolated metastatic lesion in the scalp region 15 months after treatment of her primary disease. The scalp lesion was surgically excised and radiotherapy was given. Currently, she remains disease free 4 years and 9 months following the diagnosis of her metastatic disease. Scalp metastasis from leiomyosarcoma of the parotid gland has not been reported. In fact, scalp metastasis from leiomyosarcoma of any other anatomic site or from any malignant tumor of parotid gland has not been mentioned in the literature. [Turk J Cancer 2003;33(4):191-194]

KEY WORDS:

Leiomyosarcoma, parotid gland, scalp metastasis

INTRODUCTION

Leiomyosarcoma is a malignant tumor of smooth muscle cell origin. It is most commonly found in the gastrointestinal tract and uterus. Leiomyosarcoma of the head and neck region is rarely encountered. The rarity of leiomyosarcoma in the head and neck region may be attributed to relative paucity of smooth muscle in this area. Smooth muscle is found mainly in the walls of blood vessels and the erector pili musculature of the skin (1). Presumably, leiomyosarcoma of the head and neck region originates from these structures. So far, only 8 cases of leiomyosarcoma of the parotid gland have been reported in the literature (2-7).

We report an unusual case of leiomyosarcoma of the parotid gland in a 33-year-old female who developed an isolated metastasis in the temporoparietal region of the scalp fifteen months after surgery and post-operative radiotherapy. The possible mechanisms of such metastatic involvement are also described.

CASE REPORT

A 33-year-old-woman presented in April 1996, with six months history of a slowly growing painless swelling over right side of face. She also had difficulty in closing right eye for 3-months. On examination, there was facial



Fig 1 (A,B,C,D). Clinical photographs. (1-A): showing a right facial enlargement due to parotid mass. (1-B): after parotidectomy, (1-C): scalp metastasis, (1-D): following surgery and radiotherapy for scalp metastasis

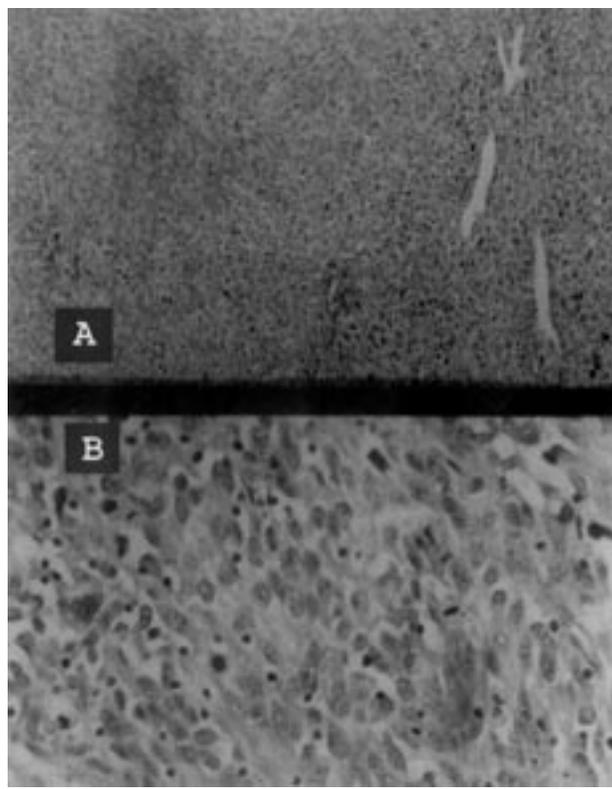


Fig 2 (A,B). Microphotographs of primary tumor (leiomyosarcoma) of parotid gland. (2-A): low power view (H&E, x10), (2-B): high power view of the same section (H&E, x40)

asymmetry due to a mass in the parotid area causing prominence of right side (Figure 1-A). The overlying skin was tense but clinically uninvolved. She had peripheral facial nerve (VIIth cranial nerve) palsy on affected side. No cervical lymphadenopathy was present.

Biopsy of the lesion was reported as poorly differentiated spindle cell sarcoma. Right parotidectomy, distal mandibulectomy and radical neck dissection was performed (Figure 1-B). Whole parotid gland was replaced by the tumor. The cut surface was grayish white, firm, nodular and measured 9.0x8.0x5.0 cm in size. The underlying mandible bone was free from the disease. Submandibular salivary gland and internal jugular vein were unremarkable. Three of the five cervical nodes at level I showed metastases. Rest of the thirteen nodes removed from level II to V were negative for metastasis. All surgical margins of resection were free. Microscopic examination (Figures 2A&B) of the parotid mass revealed elongated cells with abundant cytoplasm, marked pleomorphism and central cigar shaped nuclei arranged in fascicles. Fascicles were intersecting at right angles. Mitotic activity was noted and number of mitoses was 10-12/10 high power field. Multi-nucleated giant cells were also present. On immunohistochemical study the spindle cells showed focal positive reaction with smooth muscle specific actin, vimentin and desmin while showing negative reaction with cytokeratin (CK), AE1, epithelial membrane antigen (EMA), S-100 and skeletal muscle actin. Based on the histopathology and immunohistochemical evaluation a final diagnosis of high grade (III) leiomyosarcoma of the parotid gland was established. She received post-operative tele-cobalt therapy (total dose 60 Gy/30 Fr./6 Wk.) and remained asymptomatic till September 1997, when she again presented with a small swelling on the right side of head. On examination, it was a 1.5x1.5 cm size hard and partly fixed swelling in the right temporo-parietal region of the scalp (Figure 1-C). X-ray of the skull did not show involvement of the bone. Clinically and radiologically there was no evidence of recurrent disease at the primary site or any metastatic lesion in other parts of the body including lungs. Fine needle aspiration cytology of the scalp lesion showed malignant cells. Excision of the scalp swelling was performed. The histopathology was reported as poorly differentiated leiomyosarcoma (Figures 3A,B,C), similar to primary lesion in the parotid gland. Scalp lesion was 1.4x1.3x1.0 cm in size. Base of the resection was very close but free from the disease. Post-operatively she received radiotherapy to metastatic site

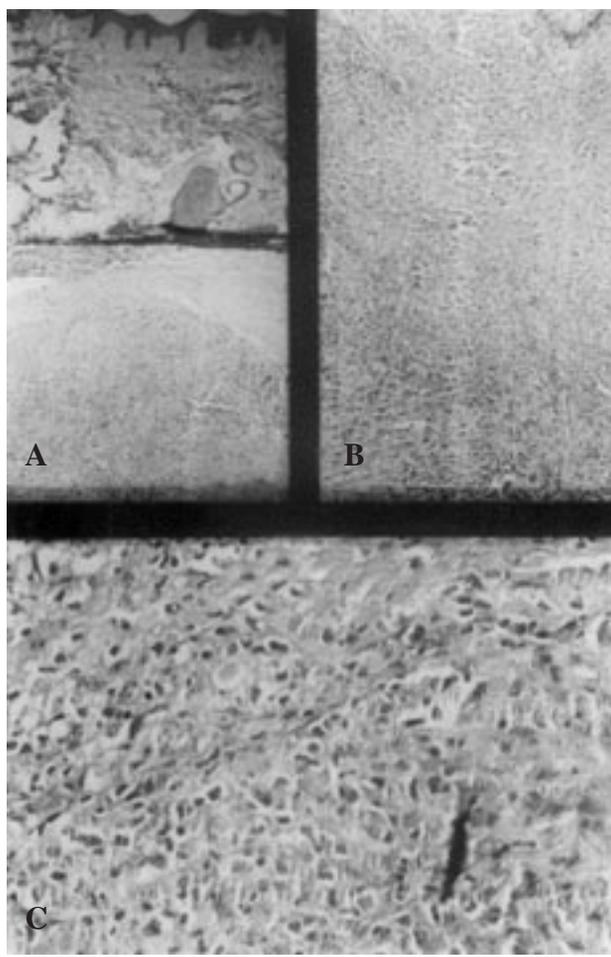


Fig 3 (A,B,C). Microphotographs of metastatic lesion in the scalp showing malignant cells diffusely infiltrating dermal connective tissue. (A): scanner view, (B): low power view (H&E, x10), (C): high power view (H&E, x40)

using 8 Mev electron beam (Figure 1-D). The patient is currently asymptomatic and without evidence of any disease 4 years and 9 months after the diagnosis of her scalp metastasis.

DISCUSSION

Soft tissue sarcomas are relatively rare tumors constituting about 0.7% of all malignant neoplasms. Leiomyosarcoma is the malignant tumor of smooth muscle cell origin. It represents only 2-8% of all sarcomas and is found most commonly in the gastrointestinal and female genital tract. Other origins such as viscera, major arteries, veins & extremities are less common sites. Leiomyosarcoma of the head and neck region is very uncommon with only a few cases at the various anatomic sites e.g. sino-nasal tract,

oral cavity, mandible, larynx, trachea, hypopharynx, cervical lymph node, external auditory canal and middle ear, etc. have been mentioned in the literature. So far, only 8 cases of leiomyosarcoma of the parotid gland have been reported (2-7).

Although the origin of leiomyosarcoma was thought to be smooth muscle cell, several authors have proposed that this tumor may, in fact be derived from pleuripotential, uncommitted mesenchymal cells or smooth muscle cells of blood vessel origin (8-12). This implies that the origin of this tumor was a small blood vessel within the parotid tissue. The tumor might also have originated in the myo-epithelial cells that are capable of multidirectional differentiation. As the whole parotid gland was replaced by the tumor, it is very difficult to point the source of origin of the tumor within the parotid gland.

Early cervical lymph node metastasis in head and neck leiomyosarcoma is rare. Leiomyosarcoma is apparently not a lesion with a high distant metastatic potential. Metastatic spread of leiomyosarcoma is usually via hematogenous route to lungs and observed in 20% of cases. Scalp metastasis, as reported in our patient has not been mentioned from leiomyosarcoma originating from any region and organ of the body or any histologic variety of a parotid malignancy in the literature. The patient developed a solitary metastasis in the scalp region 15-months after the treatment for her primary lesion. It is difficult to explain the exact mechanism of such metastatic presentation. Malignant cells reach the skin by a variety of the mechanisms including direct extension, lymphatic or hematogenous spread and through implantation in surgical scars. The so-called direct extension is spread of tumor through tissue planes, while not an actual metastasis to the skin, these tumors are included by some authors in studies of skin metastasis. The lymphatic metastases are believed to be the result of tumor cells spreading through dermal lymphatics with the subsequent deposition of the tumor cells in the skin. Distant metastasis as also seen in the present patient seems to be the result of hematogenous spread, which may involve the skin at distant site from the primary lesion. Due to normal pulmonary vascular bed filtering mechanism, distant metastases first involve the lungs in approximately 75% cases. The lungs were radiologically uninvolved in our patient. The alternative pathway could be the spread via Batson's plexus, a route of hematogenous metastases seen in various malignancies, which bypass pulmonary circulation (13). It is also possible

that, altered lymphatic drainage following initial surgery and radiotherapy for primary disease in the parotid gland could have played a role in our patient, because the metastatic site in the scalp was not very far away from the primary site in the parotid gland.

Scalp lesions may manifest clinically in several forms such as inflammatory lesions, discrete nodules or plaques. Cutaneous metastases may vary in number from a single nodule, as noted in our patient, to greater than 20 nodules in various cancers.

Cutaneous metastases including scalp lesions from various primary malignancies are rare. They are usually associated with widespread disease and carry an extremely poor prognosis. But the present patient had responded

excellently to the treatment and currently she has no evidence of disease 4 years and 9 months after the diagnosis of her scalp metastasis.

Solitary skin metastasis in the scalp region from a primary leiomyosarcoma of the parotid gland (under control), is an extremely rare feature of distant metastatic presentation in the natural history of parotid malignancy. The scalp lesion, 15-months after the treatment of primary lesion in the parotid gland could have been clinically misdiagnosed as second primary skin or another soft tissue malignancy. Proper history, thorough clinical and radiological evaluation and biopsy are necessary to establish the diagnosis.

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