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Soft tissue sarcomas of the head and neck associated with surgical trauma

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Abstract
Soft tissue sarcomas in the head and neck are rare. Aetiological factors relating to these tumours have not yet been identified. The association with von Recklinghausen's disease and with irradiation is however well recognized. In the literature it has been speculated that trauma may also play a role in the development of soft tissue sarcomas. In this article we present five patients with a history of surgical trauma at the site where a sarcoma later developed. Although we cannot prove a causal relationship, the relatively high incidence of possibly trauma-related soft tissue sarcomas in a series of 60 patients we have seen over a 30-year period, suggests that such a relationship could exist.

Key words: Head and neck neoplasms; Soft tissue neoplasms; Sarcoma; Wounds and injuries

Introduction

Soft tissue sarcomas in the head and neck are rare and account for less than one per cent of all malignant neoplasms in this region and less than 10 per cent of all soft tissue sarcomas. Known predisposing factors of soft tissue sarcomas are von Recklinghausen's disease and previous irradiation. In the literature it has been reported that scar formation and chronic infection may be predisposing factors for the development of soft tissue sarcomas (Stout, 1948; Pack and Ariel, 1952; Menkin, 1960; Gowing, 1961; Cruickshank *et al.*, 1963; Denham and Dingley, 1963; Michael and Dorfman, 1976; Hamblen and Carter, 1984; Lee *et al.*, 1984; Enzinger and Weiss, 1988; Conlon *et al.*, 1993). In a series of 60 patients with soft tissue sarcomas of the head and neck seen over a 30-year period, we identified five patients with a proven history of surgical trauma.

Case reports

In a retrospective review of clinical records over a 30-year period we identified a total of 60 adult patients who had been treated at this Institute for a soft tissue sarcoma of the head and neck. Within this group there were three patients with a history of von Recklinghausen's disease and another four patients who developed a sarcoma in a previously irradiated field. Five patients were identified who developed a sarcoma in the area of a surgical scar (see Table I). All pathological material was reviewed by one of our pathologists.

Case 1

In 1979 a 58-year-old man presented with a tumour in his left maxillary sinus. Almost 30 years previously he had undergone an antrostomy of his left maxillary sinus. One year prior to admission he complained of toothache and an impacted canine tooth was removed from his left maxilla. At that time there were also signs of numbness of the skin under the left lower eyelid, swelling and blurred vision of his left eye. Tomography showed a tumour located in the left maxillary sinus with extension into the left orbit, the ethmoid sinuses and the nasal cavity. Biopsies confirmed a malignant spindle cell tumour of mesenchymal origin.

After referral a Denker-type debulking operation of the nasosinal complex was performed, followed by radiotherapy with a total dose of 70 Gy over seven weeks. The histology showed a malignant spindle cell tumour whose morphology was consistent with a leiomyosarcoma. Immunohistochemistry for muscle-specific actin was positive while epithelial and melanoma markers were negative. Seven months later a metastasis under the left axilla was removed surgically. Two months later there was evidence of local recurrence, bone metastases and skin metastases. After two courses of palliative chemotherapy (adriamycin) there was tumour progression leading to death one year after the first visit.

Case 2

In 1980 a 70-year-old man was referred with a swelling in the upper part of the right neck (level II and V). In 1950 he had undergone a radical mastoidectomy of his right ear after which he suffered from persistent otorrhoea. One month before his referral, he developed a swelling under his right ear. An initial biopsy was inconclusive but there was a suspicion of malignancy.

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TABLE I

Patients	Age/sex	Site	Histology	Latency period in years	Therapy	Follow-up
1	58 M	Maxilla	Leiomyosarcoma	29	S + RT + CT	14 months DOD
2	70 M	Neck	Angiosarcoma	30	–	3 months DOD
3	62 M	Nose	Leiomyosarcoma	40	S + RT	7 years REC
4	46 M	Neck	Sclerosing sarcoma	8	S + RT	3 years DOD
5	69 F	Neck	Pleomorphic sarcoma	11	S + RT	15 months NED

S = surgery; RT = radiotherapy; CT = chemotherapy; NED = no evidence of disease; DOD = died of disease; REC = recurrence.

The patient presented with a diffuse peri-auricular swelling (8 cm in diameter) and in the neck several suspicious lymph nodes were felt. A CT scan showed a swelling of the soft tissues around the right external auditory canal and an infra-auricular mass. A new biopsy revealed infiltration of connective tissue by a poorly-circumscribed malignant tumour with features suggestive of an angiosarcoma. Immunohistochemistry confirmed the vascular nature of the tumour. The tumour appeared to be inoperable and was too large for radiotherapy. Since he had no complaints, no palliative treatment was given either. The tumour rapidly progressed and the patient died within three months after admission.

Case 3

In 1986 a 62-year-old man presented with right-sided nasal stuffiness, pain in his right cheek and a watering right eye. In 1945 he had undergone endonasal frontal sinus surgery. At an ENT examination a polypoid mass was seen in his right nasal cavity from which a biopsy revealed a malignant mesenchymal tumour process. CT scanning showed bony destruction of the medial wall of the right maxillary sinus by a tumour mass that completely filled the nasal cavity. A Denker-type debulking operation of the nasosinal complex was performed. Histology showed the features of a leiomyosarcoma, which was supported by the immunohistochemical findings. Post-operatively he was irradiated with a total dose of 68 Gy over six and a half weeks. There were no signs of locoregional tumour recurrence eight years after treatment. However subcutaneous tumour metastasis was diagnosed recently on the right buttock for which he underwent surgical excision.

Case 4

In 1986 a 46-year-old man presented with a clinically suspicious mass in the neck. Fifteen years before he had a car accident resulting in a 'whiplash' injury of the fifth and sixth cervical vertebrae. Because of persistent pain in his neck he was treated by repeated injections of a local anaesthetic (maeverin one per cent). Between 1977 and 1979 he received injections in the right paravertebral area at the level of C2–C6. The injections were also given around the left mastoid, near the left occipital nerve and at the right temporal area. Thirteen years after the accident he developed a mass in the left neck, at the level of C6. The mass was surgically removed from the paravertebral muscles. Histopathology revealed a malignant mesenchymal tumour, which was difficult to classify. The excision was irradical. Post-operatively he was irradiated with a total dose of 60 Gy over six weeks. One year later the tumour recurred just above the irradiated field, and for this he was admitted to our hospital.

The tumour mass was mainly located in the left suboccipital area. Incisional biopsy revealed no signs of malignancy, only scar tissue was seen with features suggestive of myositis ossificans. However the mass slowly progressed and was finally excised two years after admission. The histopathology revealed a sclerosing sarcoma with no further differentiating features. Irradiation followed with a dose of 60 Gy over six weeks. Two years later another recurrence paravertebrally at the level of C7–T3 was excised. From that time on the tumour process became rapidly progressive leading to death within one year.

Case 5

In 1992 a 69-year-old woman was referred for a soft tissue tumour in the right side of the neck. Twelve years earlier she had undergone a right hemithyroidectomy because of a nodular colloid goitre. Eleven years later she presented with a mass at the operation site, which was thought to be a recurrence of the goitre. An excision was performed using the old hemithyroidectomy scar. Surprisingly, histological examination revealed the features of a pleomorphic sarcoma. The tumour mass measured 11 × 6 × 5 cm and could not be radically excised. Several months later a recurrence of the tumour was excised with a similar histology. The patient was then referred with enlarged right neck nodes on the right side.

A radical neck dissection was performed. Histological examination revealed two soft tissue metastases in the specimen, which were excised with a narrow margin. The morphology was similar to the primary tumour. Post-operative radiotherapy was given up to a total dose of 60 Gy over six weeks. During follow-up one year after treatment there are no signs of locoregional recurrence or distant metastases.

Discussion

Little is known about the aetiology of soft tissue sarcomas. The relationship with von Recklinghausen's disease however is well recognized (Farr, 1981; Graeger and Das Gupta, 1986; Hajdu, 1993). A relatively higher incidence

of soft tissue sarcomas is also seen among hereditary syndromes, such as Turcot's syndrome, Werner's syndrome, tuberous sclerosis and multiple nevoid basal cell carcinoma syndrome (Graeger and Das Gupta, 1986), which may implicate genetic factors. In the Li–Fraumeni syndrome, a rare inherited disorder, germ line mutations of the P53 tumour suppressor gene are associated with a high risk for the development of sarcomas of bone and soft tissue, breast cancer and other tumours (Malkin *et al.*, 1990). Exposure to ionizing irradiation may also lead to the development of soft tissue sarcomas (Wilson and Brunschwig, 1937; Adam and Reif, 1977; Donaldson, 1978; Farr, 1981; Graeger and Das Gupta, 1986; Enzinger and Weiss, 1988). Furthermore, a relationship has been documented between chronic lymphatic stasis after radical mastectomy and lymphangiosarcoma (Stewart–Treves syndrome) (Gowing, 1961; Laskas *et al.*, 1975; Conlon *et al.*, 1993). Since fibroblasts are actively involved in all types of repair processes, it has been speculated that trauma may act as a causative factor in the aetiology of soft tissue sarcomas. In this respect the development of fibrosarcomas in scar tissue showing fibroblastic proliferation is not surprising (Stout, 1948; Heller and Sieber, 1950; Ivins *et al.*, 1950; Pack and Ariel, 1952; Swain *et al.*, 1974; Enzinger and Weiss, 1988).

When a malignant tumour originates at the site of previous trauma the question is whether the association is coincidental or represents a true cause-and-effect relationship. The low incidence of malignant tumours compared with the high incidence of different types of trauma, including surgical trauma, suggests that the risk of inducing a neoplasm is minimal at most. Nevertheless trauma-induced soft tissue sarcomas have been widely reported in the literature (Stout, 1948; Heller and Sieber, 1950; Ivins *et al.*, 1950; Pack and Ariel, 1952; Swain *et al.*, 1974; Enzinger and Weiss, 1988).

With respect to our five patients a causative relationship between trauma and sarcoma formation cannot be proven. However, the relatively high incidence of surgical trauma in our series of 60 patients with soft tissue sarcomas is surprising, and is not seen in patients with, for example, squamous cell carcinoma. Since the injuries reported in the clinical records are surgical, and not a coincidental finding, an aetiological relationship may be more likely. Moreover, the latency periods found in our cases are in keeping with those reported in the literature (Waugh, 1952; Gowing, 1961; Denham and Dingley, 1963; Weinbren *et al.*, 1978; Enzinger and Weiss, 1988; Jennings *et al.*, 1988).

Fibrosarcomas have mainly been reported as being related to scar formation (Stout, 1948; Heller and Sieber, 1950; Ivins *et al.*, 1950; Pack and Ariel, 1952; Swain *et al.*, 1974; Enzinger and Weiss, 1988). More recently there have been reports of several cases of sarcomas associated with metal implants or foreign body material and sarcomas following an intramuscular injection (Burns *et al.*, 1972; Lee *et al.*, 1977; Weinbren *et al.*, 1978; Tayton, 1980; Lee *et al.*, 1984; Penman and Ring, 1984; Weber, 1986; Hughes *et al.*, 1987; Jennings *et al.*, 1988; Lamovec *et al.*, 1988; Conlon *et al.*, 1993). These sarcomas had a variety of histological diagnoses. In our patients no fibrosaromas were found. Remarkably, we found two cases of leiomyosarcoma with an almost identical history of paranasal sinus surgery.

In *Cases 1, 2* and *3* chronic infection and bony destruction appeared to play a significant role. Some authors have suggested a relationship between chronic reparative processes and the development of malignancies (Golberg, 1960; Gowing, 1961; Cruickshank *et al.*, 1963; Michael and Dorfman, 1976; Lee *et al.*, 1984). Furthermore an association of sarcomas with chronic draining osteomyelitis is well documented (Waugh, 1952; Cruickshank *et al.*, 1963; Denham and Dingley, 1963; Morris and Lucas, 1964). Gowing (1961) hypothesized that the tumour is often preceded by chronic and severe scarring leading to a disturbance of normal tissue relationships. He suggested that it is therefore possible that tumours may develop in tissues which are the seat of extensive disorganization, scarring and chronic inflammation. Michael and Dorfman (1976) described the relationship of malignant fibrous histiocytoma to bone infarcts and suggested that chronic reparative and proliferative changes around bone infarcts predispose to the development of malignant lesions.

The operations in *Cases 1, 2* and *3* involved bony destruction and certainly caused areas of bone necrosis. This, combined with chronic inflammation, may have stimulated a chronic reparative process which might have played a role in the development of a sarcoma 29, 30 and 40 years later respectively. In *Case 4* the patient received numerous injections with local anaesthetic (maeverin) at the site of his subsequent sarcoma. Several articles have been published about sarcomas originating at injection sites (Golberg, 1960; Greenberg, 1976; Lee *et al.*, 1977; Weinbren *et al.*, 1978; Jennings *et al.*, 1988). In this respect the injection of iron dextran has been mentioned as a causative factor. Golberg (1960) comments on this by suggesting that these sarcomas are probably not caused by iron particles but that repeated trauma followed by muscle necrosis and chronic inflammatory changes caused by injections of a wide variety of chemical agents may lead to the production of sarcomas. A sarcoma following an intramuscular injection of penicillin has also been described (Lee *et al.*, 1977). The author suggests that malignant transformation may arise in a haematoma or myositis ossificans following the injection.

Conclusions

We have presented five cases of soft tissue sarcomas originating at the site of previous surgical trauma in the head and neck. Although these sarcomas originated at the site of surgical trauma after a long latency period, a real cause-and-effect relationship cannot be proven. The rarity of tumours in scars seems to support the suggestion that trauma is no more than a co-carcinogenic factor and that additional factors are necessary for tumour formation.

Since the best hope for survival lies in early diagnosis, it is necessary to be alert to the possibility of a sarcoma when a mass arises at the site of former surgical trauma.

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