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Review Article

Aggressive fibromatosis of the head and neck (desmoid tumours)

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Abstract
Desmoid tumours are histologically benign fibrous neoplasms arising from the musculoaponeurotic structures throughout the body. They are characterized as infiltrative, usually well-differentiated firm overgrowths of fibrous tissue and are locally aggressive. An aggressive clinical behaviour with tendency for recurrence makes the treatment of these relatively rare fibrous tumours difficult. Local recurrence rates are reported as high as 70 per cent of cases, the recurrence of desmoid fibromatosis in the head and neck is difficult to ascertain because of the different classification schemes used by different authors. A review of cases of desmoid tumours of the head and neck presented in the last 10 years is reported in this paper. Complete surgical excision of desmoid tumours is considered to be the only effective method of cure by most authorities.

Key words: Fibromatosis, Aggressive; Head and Neck Neoplasms

Introduction

Desmoid tumours are histologically benign fibrous neoplasms arising from the musculoaponeurotic structures throughout the body. Extra-abdominal desmoid tumours account for almost a third of all desmoid tumours, occurring most frequently in the shoulder or thigh regions.[1] Head and neck desmoids comprise only 11 to 15 per cent of extra-abdominal desmoids. Fibrous tissue tumours of the nasal cavity, paranasal sinuses and nasopharynx are rare.

Conley,[2] reported 40 cases with fibromatosis of the head and neck, four of which were in the nasal cavity and paranasal sinuses. Among 256 patients with non-epithelial neoplasms involving the nasal cavity, paranasal sinuses and nasopharynx, only six cases were of a desmoid nature.[3] Only five cases of sino-orbital desmoid fibromatosis have been reported in the literature so far, with four involving paediatric patients.[4] They are characterized as infiltrative, usually well-differentiated, firm overgrowths of fibrous tissue and locally aggressive,[5] with a propensity to invade and erode bone and soft tissues, putting vital structures in the head and neck region at risk.[6,7]

An aggressive clinical behaviour with a tendency for recurrence makes the treatment of these relatively rare fibrous tumours difficult. Local recurrence rates are reported as high as 70 per cent of cases.[8] The recurrence of desmoid fibromatosis in the head and neck is difficult to ascertain because of the different classification schemes used by different authors.[9]

Patients and methods

A review of cases of desmoid tumours of the head and neck presented in the last 10 years to two ENT departments is reported in this paper. The head and neck region is defined as that portion of the body above the clavicle anteriorly, superior to the first thoracic vertebral spine posteriorly, and above the should girdles laterally.

Histological diagnosis of desmoid tumours and a minimum follow-up period of two years were necessary for inclusion in the study.

The common congenital tumour of the sternocleidomastoid muscle of infants was excluded. All pertinent data were abstracted from the records. Particular attention was directed to the initial treatment of the tumour, recurrence, and subsequent therapy.

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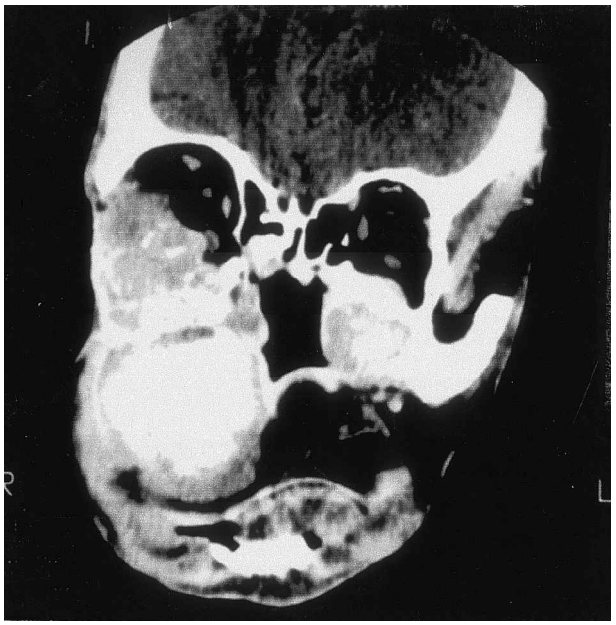

FIG. 1
Coronal CT scan of the paranasal sinuses of *Case 1*.

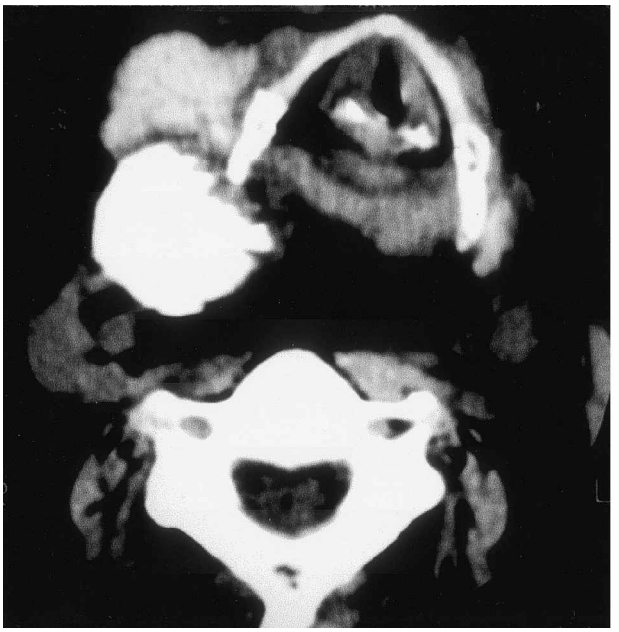

FIG. 3
Axial CT scan of the neck of *Case 3*.

Case 1

A 29-year-old female presented with a 12-month history of facial pain followed by diplopia and right-sided facial swelling. On examination she had a right-sided facial non-tender swelling with frontolateral right proptosis. Computed tomography (CT) scan (Figure 1) showed an extensive locally destructive lesion involving the right maxillary sinus, orbit, pterygopalatine and infra-temporal fossae.

The patient had had a right radical antrostomy 10 months previously assuming simple sinus disease, pathological confirmation of the type of tumour was carried out by both gingival and antral biopsies. Midfacial degloving and total right maxillectomy including clearing both the pterygopalatine and infra-temporal fossae, were followed by chemotherapy.

Local recurrence occurred 18 months later and death followed eight months later from the extensive local destructive recurrence and bleeding.

Case 2

A 49-year-old male presented with stridor, he had a neck swelling that had been neglected for 18 months and progressive dysphagia. He reported two previous incisional biopsies. CT neck (Figure 2) showed

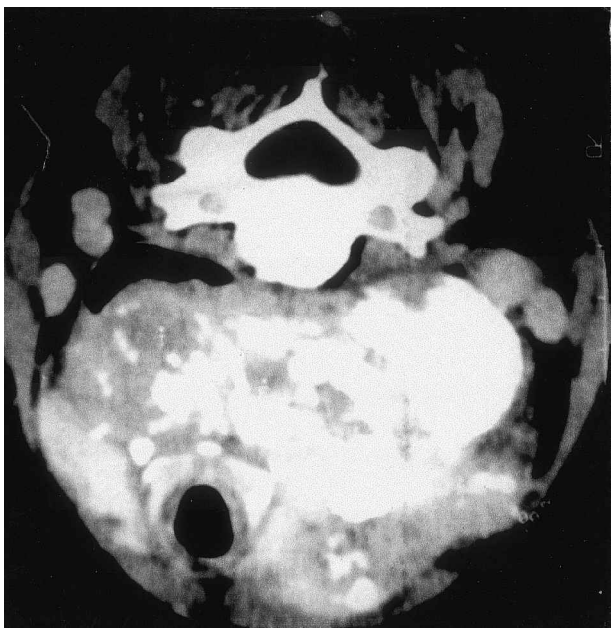

FIG. 2
Axial CT scan of the neck of *Case 2*.

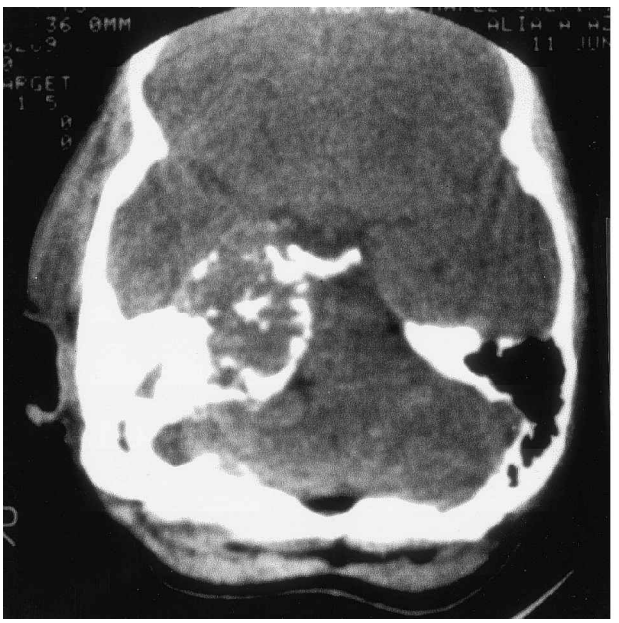

FIG. 4
Axial CT scan of the brain of *Case 4*.

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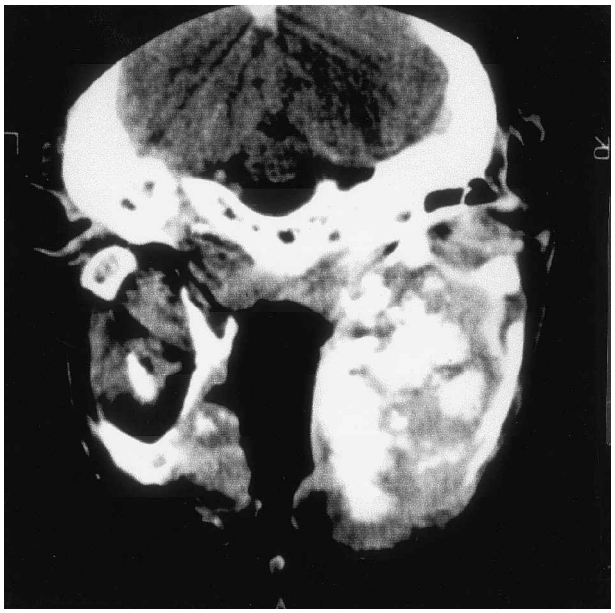

FIG. 5
Coronal CT scan of the paranasal sinuses of *Case 5*.

extensive bilateral cervical involvement of a destructive mass.

Urgent tracheostomy was performed, followed by planned right radical neck dissection and left functional dissection, there was histological confirmation of the nature of the tumour. He was given chemotherapy but there was local recurrence six months later and he died seven months later from a carotid blow out.

Case 3

A 31-year-old female presented with a 16-month history of an asymptomatic right neck mass. Three months previously she had developed progressive dysphagia. She had undergone tonsillectomy and cervical lymph node biopsy previously for the establishment of a diagnosis, CT neck (Figure 3) showed a right parapharyngeal mass. Planned right radical neck dissection was performed following histological confirmation by fine needle aspiration cytology (FNAC), followed by DXT and chemotherapy.

Repeated follow-up up to 40 months following her surgery showed no evidence of recurrence.

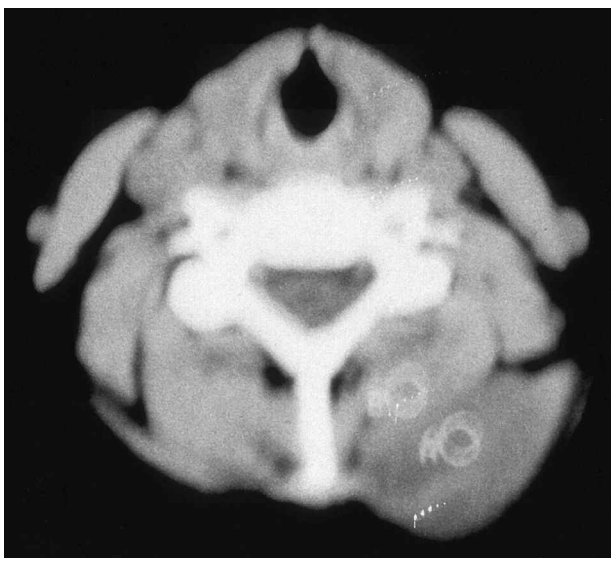

FIG. 6
Axial CT scan of the neck of *Case 6*.

Case 4

A 42-year-old female presented with trigeminal neuralgia symptoms, right-sided hearing loss and diplopia of eight months duration, that were resistant to usual neuralgia medications. She had right abducent nerve paralysis. CT scan (Figure 4) revealed a mass in the right petrous apex. Planned subtotal petrosectomy was performed. Twenty-one months later there was no evidence of recurrence.

Case 5

A 42-year-old male presented with trismus and a right temple mass of a few months duration. CT scan (Figure 5) showed a mass involving the right temporal fossa down to the infra-temporal fossa. Complete mass excision by the lateral skull base approach was performed, followed by chemotherapy. Recurrence six months later was followed by re-excision. There was no evidence of recurrence 12 months later.

Case 6

A 32-year-old female presented with left shoulder pain and swelling in the posterior triangle of the neck. On examination the swelling was well-defined, 4 cm in diameter and deep to the trapezius muscle as confirmed by CT scanning (Figure 6). FNAC revealed the type of tumour, it was excised completely with free histological margins, and there was no evidence of recurrence at 25-month follow-up.

Discussion

Historically, the term 'desmoid' refers to the hard, tendon appearance of the tumour. Bennett[10] described a group of tumours that he called 'fibronucleated cancroid growths'. His description and comments concerning three such tumours occurring on the thigh, in the parotid region, and on the arm, respectively, suggest that these may have been desmoid tumours.

Lesions occurring in extra-abdominal locations were usually classified as fibrosarcomas. During the past two decades a marked re-surgence of interest in the tumours of fibrous tissue origin has occurred.

Gross and histologic features

A desmoid tumour may be defined as an aggressive form of fibromatosis that presents as a circumscribed, locally infiltrative, well differentiated, firm

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overgrowth of fibrous tissue arising in the musculoaponeurotic structures of the body.[11]

The tumour has the gross appearance of interlacing trabeculae of mature fibrous tissue with a propensity to infiltrate surrounding muscle, adipose tissue, and fibrous tissue. It encases blood vessels and nerves without apparent deep invasion of these structures. Periosteal invasion and bone erosion are occasionally encountered. Histologically, the tumour is composed of mature fibroblasts with a generous collagenous component[12] In general, desmoid tumours are rare, with an estimated incidence of two to four cases per million patients per year.[13–14]

Desmoid tumours of the head and neck are extremely uncommon tumours associated with a high recurrence rate similar to that for extra-abdominal desmoid tumours, and estimated to recur in 25 to 70 per cent of cases[15] A total of 82 cases of head and neck desmoids (excluding our current series) have been reported in the literature.[16] Metastasis does not occur, although degeneration into malignancy has been reported.[17] Although death secondary to desmoid tumours is unusual, it has been reported in six cases.[18] Distant metastasis from desmoid tumours has not been observed in our series nor, to our knowledge, has it been reported in the literature. Death from local extension into vital structures is common and malignant changes are rare.[19]

On CT scans (Figures 1–6), the lesion is usually hyperattenuating (44–49 Hounsfield units), enhancing significantly after application of contrast material (63–66 Hounsfield units). The magnetic resonance image (MRI) shows a multilobulated lesion of heterogeneous signal intensity. The tumour is markedly hypointense on T2-weighted images and slightly hypointense on T1-weighted images relative to brain tissue, iso- or slightly hyperintense relative to tongue muscle on both T2- and T1-weighted images.[20]

Incidences of trauma have been reported to precede extra-abdominal desmoid tumours in 19 to 49 per cent of patients. Previous attempts at incision, blunt injury, and even the contractions of pregnancy and delivery have been theorized to play a role in an abnormal response to healing with persistent immature fibroblast formation giving rise to the tumour.[21]

A hereditary propensity toward abnormal connective tissue repair incited by trauma and growth by steroid sex hormones have been proposed.[22]

Recurrence was most common in patients with supraclavicular or neck masses. Complete surgical excision of desmoid tumours is considered to be the only effective method of cure by most authorities.[23]

Radiotherapy for desmoid tumours has been employed in the past either alone or in conjunction with surgical excision with little success.[24]

Although no definite benefit could be attributed to irradiation, it did appear that in several cases, irradiation may result in a transient decrease in tumour size and consistency.[25] Other treatments have included prednisolone, antioestrogens, castration, theophylline, indomethacin or vitamin C.[26]

Generally, the effect of such alternatives has been sporadic or transient. Although the majority of recurrences occur within 18 months of initial treatment, all patients should have long-term follow-up, as recurrences have been reported 12 years from the primary procedure. The treatment of recurrences should be aggressively pursued with excision of lesions amenable to extirpation without sacrifice of major structures if possible.

Conclusion

Desmoid tumours are histologically benign fibrous neoplasms. They are characterized as infiltrative, usually well-differentiated firm overgrowths of fibrous tissue and are locally aggressive. Treatment consists of an adequate three-dimensional imaging evaluation, followed by an en bloc resection with adequate margins. Post-operative radiotherapy, chemotherapy, and recently, hormonal adjuvant therapy have been used in an effort to control local recurrence.

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