Case 18

A 39-year-old Thai man from Nonthaburi

Present history:

Chief complaint: Slow growing mass at forehead for 15 years

Present illness: The patient noticed asymptomatic mass at right temporal area for 15 years. During last five year it gradually increased in size, subsequently he developed a new small slow growing nodule adjacent to the old one for 6month. He had no other systemic symptom except loss of appetite and lose his weight about 10 kilograms within 1 year.

Past history:

Myasthenia gravis S/P thymectomy on prednisolone 10 mg/day

Asthma

Family history:

Nil

Physical examination:

Vital signs: Normal

Skin: 2 well-circumscribed, rubbery to firm, erythematous nodules at right temporal area, diameter 0.5 cm and 3 cm.

Lymph node: can't be palpated

Liver and spleen can't be palpated

Neurological examination: intact

Investigation :

Skull plain x-ray shows irregular radiolucencies line along right temporal area.

CT brain: pending

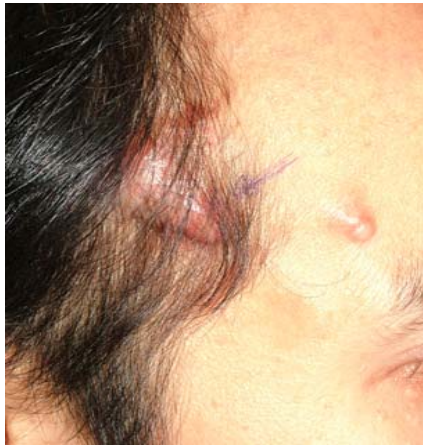

Fig. 18.1

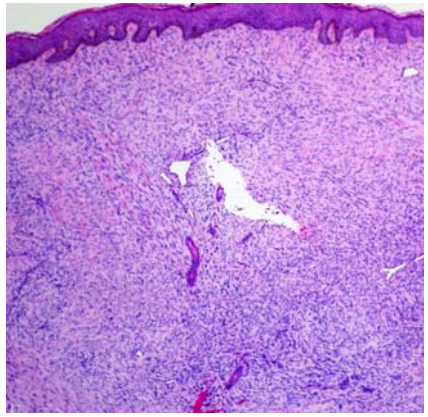

Fig. 18.2

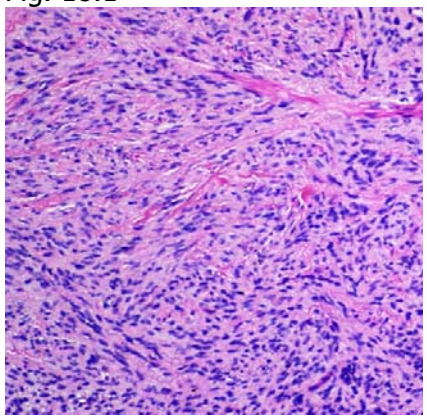

Fig. 18.3

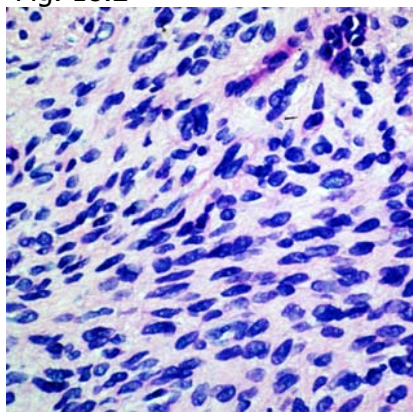

Fig. 18.4

Histopathology (s09-007338) (Fig. 18.2-18.4)

- A dense diffuse proliferation of uniform plump spindle cells in the entire dermis
- Some tumor cell arranged in short fascicles and some in storiform pattern
- Normal epidermis

Diagnosis : Dermatofibrosarcoma protuberans (DFSP)
Presenter : Chayada Kokpol
Consultants : Ploysyne Busaracome

Discussion:

Dermatofibrosarcoma protuberans (DFSP) is a locally aggressive sarcoma of intermediate malignancy that typically occurs during the third and fourth decades of life, although it has been reported in patients from infancy to ninth decade.[1]

DFSP accounts for less than 0.1% of all malignant neoplasm and represents 2-6% of all soft tissue sarcomas. DFSP occurs on the trunk in 50-60% of patients, the proximal extremities in 20-30%, and the head and neck in 10-15%. There is a predilection for the tumor to involve the shoulder or pelvic areas.[1,2]

The pathogenesis is not fully understood. Recent genetic research has been able to show chromosomal translocations or ring chromosomes which occur in DFSP through a fusion of the chromosome regions 17q22 and 22q13. Such that the collagen Type Iα1 gene (*COL1A1*) becomes fused to the platelet-derived growth factor (PDGF) β-chain gene (*PDGFβ)*. This rearrangement results in the deregulation of PDGF β-chain expression and leads to continuous activation of the PDGF receptor β (PDGFRβ) protein tyrosine kinase, which promotes DFSP cell growth.[3,4]

The clinical morphology of DFSP is variable. The most common presentation is a slow growing; asymptomatic, skin-colored indurated plaque that eventually develops violaceous to red-brown nodules measuring from one to several centimeters in diameter. On palpation the lesion is firm and attached to the subcutaneous tissue. It also can present as a nonprotuberant, atrophic, violaceous lesion resembling morphea, sclerosing basal cell carcinoma, anetoderma, or scar.

The clinical differential diagnosis includes keloid, large dermatofibroma, dermatomyofibroma and morphea.[1,2]

Pathologically, DFSP is composed of fascicles of densely packed, monomorphous spindle cells arranged in a storiform (mat-like) pattern. Lesions are poorly circumscribed, with diffuse infiltration of dermis and subcutis yielding a honeycomb pattern. Infiltration into the underlying fascia and muscle occurs as a late event. In less than 5% of cases a DFSP may contain melanin-producing cells with schwannian differentiation and this is termed a Bednar's tumor (pigmented DFSP).[1,3]

The general immunostaining pattern of DFSP – CD34-positive, factor XIIIa-negative – serve to distinguish it from large and/or highly cellular dermatofibromas, which are CD34-negative, factor XIIIa- positive. It has also been shown that dermatofibromas strongly express CD44 with only faint stromal hyaluronate, where as DFSP shows reduced or absent CD44 and strong stromal hyaluronate deposition.[5]

In the international classification of soft part tumors, DFSP is allocated to the group of fibrohistiocytic tumor with intermediate malignancy. There are no universally accepted stages. A simple division into 3 stages can be used: Stage I: primary tumor alone; Stage II: locoregional recurrence; Stage III: remote metastasis. It will possibly be most practical in future to subdivide Stage I into Ia (without sarcomatous parts) and Ib (with sarcomatous parts).[5]

Tumor extent and mobility generally are assessed on physical examination. DFSP rarely exhibit lymphatic or hematogenous dissemination; regional lymph nodes are assessed by palpation. Magnetic resonance imaging (MRI) is useful for ascertaining deep tumor invasion, particularly in patients with large recurrent lesions.[5]

Surgical removal with clear margins is the goal with DFSP. Approximately 30-50% of DFSP recur locally after simple excision, so it generally recommends that a wide excision with margins of 1 to 3 cm should be performed. Risk of local recurrence decreases with increasing surgical margins. Multiple case series have shown Mohs

microscopic surgery to be an extremely effective method of resection of DFSP, with very low rate of local recurrence.[1,2] Because of the ability to minimize surgical margins, Mohs microscopic surgery is considered as the treatment of choice in particularly anatomically challenging areas such as head or neck and in the treatment of children.[6,7]

Adjuvant radiation therapy may help decrease the local recurrence rate. The use of imatinib, a chemotherapeutic agent that target the molecular translocation distinctive to this tumor, has shown promise in this setting of localized advanced disease or metastatic disease. Imatinib may also emerge in the future as an adjunct to surgery.[2]

References

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