Morphea Syndrome

Morphea is a rare connective tissue disorder with unknown etiology. It is a form of scleroderma which is characterized by cutaneous sclerosis and systemic involvement. Sclerosis can be of two types: first is systemic sclerosis with skin involvement and visceral manifestation and second is localized scleroderma which is also known as morphea with cutaneous manifestation with/without involvement of underlying tissues. It is a benign condition. Morphea should be treated early as its complications can cause high morbidity.

KEYWORDS: morphea syndrome, scleroderma, en coup de sabre.

INTRODUCTION

Morphea also known as localized scleroderma is characterized by sclerosis of the skin and in underlying tissues in some cases. Currently it is divided into categories based solely on cutaneous morphology, without taking into account the systemic disease or autoimmune phenomena. Autoimmune basis of morphea is suggested by presence of phenomena such as vitiligo and Hashimoto thyroiditis with morphea in some cases. Morphea is differentiated from systemic sclerosis by the absence of vasospasm, structural vascular damage, and involvement of internal organs, and also by the distribution of the skin lesions.
CASE REPORT
A Female of 31 years of age completely asymptomatic before 2 years, developed a marble sized, hard, lump in her right breast upper outer quadrant with overlying skin black in colour without pain. On USG report it was diagnosed as focal duct ectasia. Then within next few days the size and hardness of lump gradually increased. On X-ray nothing specific was found. But instead of resolving within few days the lesion was gradually increasing.

Then biopsy of the lesion was done. The report of biopsy suggested that it could be either dermatitis or collagen disease. The microscopic features on biopsy suggested that epidermis is atrophic, no evidence of spongiosis, dermis shows increased collagen, collagen is disorganised and there is perivascular inflammatory infiltrates and no fibrinoid necrosis. After this report she was suspected to be having collagen vascular disease and was on treatment for the same. On general examination her bloodpressure, blood sugar were normal. She has no family history of similar complains. On other investigations like CBC, ESR, Urine routine, SGOT, SGPT, Rheumatoid factor, Platelet count, ANA (Immunofluoroscence) nothing significant was found. She did not respond to the treatment for collagen vascular disease and her lesion kept on increasing and similar lesions appeared over left upper limb.

On second biopsy she was diagnosed as having morphea syndrome and was referred to rheumatologist. Then she was started on treatment which included Tab. Folitrax (methotrexate), Tab. folvite (folic acid), Tab. akilos-p (Aceclofenac and paracetamol), capsule Essential (polyenylphosphatidylcholine) and tab. rekool (rabeprazole) for morphea syndrome by her rheumatologist and she responded well to it. Her lesions gradually started resolving. She is right now under medication and on regular follow up. Her lesions has now subsided but light scar marks are present.

DISCUSSION
Morphea is a connective tissue disorder of unknown etiology or also known as localized scleroderma. Two categories of scleroderma are known: systemic sclerosis (SSc), which is characterized by cutaneous sclerosis and visceral involvement (especially the esophagus, lung and vascular system); and localized scleroderma (LoS), which classically presents benign and self-limited evolution which is confined to the skin and/or underlying tissues.\(^3\) Localised scleroderma or morphea is a rare disease and has an incidence of around 0.3 to 3 cases per 100,000 inhabitants/year.\(^4\) In our case the patient has localised scleroderma. The characterestic feature of Morphea is skin thickening with increased quantities of collagen in
indurative lesions.[5] Some studies have indicated an association of autoimmune diseases with morphea and also an increased frequency of autoantibodies in serum with morphea.[6]

CLASSIFICATION OF MORPHEA: The most widely used classification in the literature is the Mayo Clinic Classification (in its simplified form) According to this classification 5 types of morphea are as follows.[7]

1. Plaque morphea,
2. Generalized morphea,
3. Bullous morphea,
4. Linear scleroderma - including subtypes that involve the head and face, linear scleroderma ‘encoup de saber’ (LScs) and progressive facial hemiatrophy (PFH),
5. Deep morphea.

In our case the patient has plaque morphea. The plaque morphea is characterized by limited, round or oval shaped areas of hard and shiny skin, and it affects one or more anatomical area, mostly the trunk and proximal extremities are affected. In the earliest phases, it presents with violaceous halo can be seen around the plaque(“purple ring”); this corresponds to the inflammatory phase of morphea.Bullous morphea which is a rare form of morphea is mainly characterized by the appearance of bullae or erosions on plaques.[8]

In deep morphea the overlying skin may be normal or atrophic or hardened, and will almost always be depressed or adhered to the deep plane. It is usually asymptomatic and usually not associated with involvement of viscera.[9, 10] Generalized morphea is characterised by morphea plaques involving more than 2 body sites. It is more commonly seen in women, and physical exercise has been identified as a triggering factor. The plaques in this are slightly inflamed, pigmented, ill-defined, thickened, adhered to deep planes, fascia and muscle and this is mostly seen over the trunk and extremities.Onset of sclerosis is gradual and then relatively fast over a period of months.In this signs of acute inflammation such as edema and erythema may not be present.[11] Linear scleroderma is also known as “en coup de sabre” (LScs) which is a rare and intriguing form of LoS, which was first described by Addison in 1854.[12] Etiology of morphea includes genetic factors.[13] Vascular abnormalities occurring in scleroderma have also been reported as an etiological factor.[14]
The treatment of morphea is still unsatisfactory and there are very few randomized and controlled therapeutic studies.\textsuperscript{[15]} Various therapeutic modalities have been suggested which includes the use of topica l medications, immunosuppressive pharmacological agents, physical therapy and phototherapy.\textsuperscript{[16]}

**CONCLUSION**

MORPHEA is not an exclusively cutaneous disease. During the diagnosis core attention needs to be paid on its association with other connective tissue diseases. Sometimes transitional form of morphea may be present. Management of morphea is a bit challenging and evaluation is difficult as sclerotic lesions can resolve spontaneously sometimes.

**REFERENCES**


