Journal of Pakistan Association of Dermatologists. 2018; **28(4)**: 395-397.

Editorial

Screening for esophageal involvement in morphea: Is it needed?

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Morphea, also known as localised scleroderma (LS), is a chronic inflammatory disease characterized by sclerosis of the skin due to excessive collagen deposition in the dermis and subcutaneous tissue. It is clinically characterized by round or oval, irregular or linear plaques that are initially dull red or violaceous, smooth and indurated but later turn atrophic and histologically present with sclerosis of the dermis and/or subcutaneous tissue. The internal organ involvement in morphea is usually not seen.[1,2] Systemic sclerosis (SSc) on the other hand is a generalized/systemic form of scleroderma affecting the skin and various organ systems of the body like gastrointestinal tract, lungs, heart and kidneys. About 90% patients of SSc have gastrointestinal tract involvement, of which esophagus is the most frequently affected part. In SSc, esophageal smooth muscle becomes atrophied and later replaced by fibrous tissue that causes severe disturbances in the motility of distal esophagus manifesting as reduced lower esophageal sphincter pressure (LESP) and loss of distal esophageal body peristalsis.[2-4] Thus the esophageal involvement in SSc is well known and has been studied extensively. However, the gastroesophageal involvement in morphea has been studied scarcely. While most studies are against the gastroesophageal involvement in morphea, a handful of studies have revealed some mild and infrequent involvement of the same.

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Medical literature is very deficient in the studies where gastroesophageal tract has been studied in patients of morphea. Zaninotto *et al.*[3] studied esophageal motility and LESP in 13 patients of SSc and 16 patients of LS by means of esophageal manometry and 24-hour pH monitoring of the distal esophagus. Marked abnormalities in esophageal motility and in acid exposure in the distal esophagus were observed in SSc patients only. Their data suggested that marked esophageal involvement is present only in the systemic form of scleroderma i.e. SSc. They recommended that esophageal tests may be useful in the evaluation of SSc; however, their use does not seem to be justified as routine in patients of LS.[3]

Weihrauch and Korting[4] studied esophageal function by radiography and manometry in 51 patients of SSc, 14 patients with morphea, 12 patients with Raynaud's disease and 21 normal control subjects. Peristaltic contractions of the lower two-thirds of esophagus were affected only in SSc. LESP was significantly decreased only in the patients of SSc but was normal in morphea and Raynaud's disease.[4]

Dehen *et al.*[5] studied 76 patients with morphea and found internal involvement by systematic examination in 16 patients only. Out of these 16 patients, two (including one patient who

developed SSc) had symptomatic and severe visceral disease. However, the other 14 had asymptomatic and minor abnormalities consisting of abnormal LESP and/or peristaltic abnormalities in the esophagus. After a median follow-up of 4 years, such involvement did not affect the prognosis in these patients. The mildness of these visceral abnormalities suggested that the routine screening of these asymptomatic patients is not justified. They concluded that morphea and SSc behave as two different diseases.[5]

Recently, the author and associates have carried out a prospective study in which esophageal involvement was investigated in 56 patients of SSc and 31 patients of morphea by clinical, endoscopic, manometric and pH metric features. Reflux esophagitis was seen in 17 (32.7%) cases of SSc and only two (7.14%) cases of morphea. Manometric abnormalities were seen in 32 (68.1%) cases of SSc and none in morphea. Ambulatory 24-hour esophageal pH monitoring documented abnormal reflux in 33 (80.5%) of SSc cases and no such abnormality was seen in morphea. It was concluded that though esophageal involvement is frequent in SSc, no such motility disorder is seen in morphea. It was suggested that meticulous upper gastrointestinal tract evaluation is justified only in patients of SSc and not in morphea.[2]

Callen[6] at the NEJM journal watch, reviewed the esophageal involvement in morphea and concluded that esophageal evaluation need not to be carried out in the patients of morphea.[6]

Though it seems that gastrointestinal involvement is not seen in patients of morphea; however, there are a very few studies which have revealed esophageal involvement in morphea regardless of the type of morphea. At this point of time it will be pertinent to mention them here. Guariso *et al*.[7] evaluated the esophageal involvement in 14 patients with juvenile LS. Esophageal abnormalities were found in 8/14 (57%) patients: pathological acid exposure on 24-hour pH monitoring was found in 7; non-specific esophageal motor abnormalities in 5 and endoscopy-proved esophagitis in 5 symptomatic patients. Interestingly, 5 out of 8 patients with esophageal abnormalities were found to be ANA positive, and 2 were also RF positive. The high percentage of esophageal involvement in this study can be due to the very low number of patients. The authors concluded that esophageal involvement is not unusual in patients with juvenile LS, especially in presence of positive autoantibodies or specific GI symptoms.[7] Similarly, a review done by Zulian *et al*.[8] comprising 750 children with LS allocated from 70 European centres showed that gastrointestinal involvement was seen in 6% of the patients.[8]

Thus summing all together, it can be inferred that, most studies have not revealed any significant and frequent involvement of upper gastroesophageal tract in patients of morphea/ LS. However, from the current knowledge of the subject, a complete absence of esophageal involvement in LS especially juvenile LS cannot be ruled out. Hence routine screening for esophageal involvement in patients of LS is not justified. However, patients of juvenile LS who are symptomatic for gastroesophageal symptoms or having autoantibodies can be subjected to meticulous history regarding gastroesophageal symptoms and referral to a gastroenterologist can be considered based on patient's clinical profile.

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