Dysphagia Due to Forestier Disease: Three Cases and Systematic Literature Review

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Abstract Forestier disease or diffuse idiopathic skeletal hyperostosis (DISH) is an uncommon cause of dysphagia. Due to rarity of this condition there is neither any demographic data nor any consensus about the investigation and treatment modalities. Here an effort is made in the present article to compile the information regarding the demographic and clinical features, investigation modalities and

different methods of treatment from all the available case reports of dysphagia due to Forestier disease in the English literature till date. Three cases of the same condition are also reported in the present paper. Three cases of dysphagia due to Forestier disease treated in the ENT department of R.G. Kar Medical College and Hospital are reported herewith. A systematic review of literature is also done. All the case reports of dysphagia due to Forestier disease are obtained by World Wide Web search (WWWS) using ‘Forestier Disease’, ‘DISH’ and ‘dysphagia’ as the key words. Data regarding age, sex, duration of the symptom, associated symptoms, investigations done, level of vertebrae involved and different modalities of treatment with result are collected and analyzed systematically. We found total 73 cases of dysphagia due to Forestier disease. The condition commonly affects males (M:F 5.64:1) in older age group (94.52% cases are 60 years or above), often presented to the treatment facility after a long time of initiation of the symptom. Barium swallow X-rays and lateral neck X-ray were the most common investigations done to arrive at a diagnosis as per result of systematic review. Surgical removal of the causative osteophytes were the definitive treatment, but if surgery is contraindicated, conservative measures like switchover to liquid and semi-solid food and proper swallow training also improve the condition to some extent. Dysphagia due to Forestier disease mostly affects older male, often has a chronic course. It can be diagnosed with simple investigations like neck X-ray or barium swallow X-rays. The definitive treatment is surgical, but if contraindicated the problem can be palliated with simple measures like swallow training and change of food.

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Introduction

Dysphagia is a common presenting symptom in otolaryngology clinics all over the world. There is a variety of etiologies for this symptom, among which anatomical narrowing of the esophageal lumen is the most important. Narrowing may be due to tumors, abscesses etc. originating from the esophagus itself or from the surrounding area. Diffuse idiopathic skeletal hyperostosis (DISH) involving the cervical spine is a rare cause responsible for this distressing symptom. Forestier and Rotes-Querol in 1950 first described the condition and separated the disease from discarthrosis and ankylosing spondylosis [1]. After the name of the pioneer the condition is also named as Forestier disease. Due to the rarity, there are only a few case reports and series of this condition and there is no study found in English literature after extensive search. In the present study we report three cases of Forestier disease, presented at the outpatient department of R.G.Kar Medical College & Hospital, Kolkata, India within last 10 years. An effort is also made in the present study for a systematic review of the demographic data and trend for investigating and treating the condition reported in different case reports and serie till date through World Wide Web Search (WWWS).

Materials and Methods

Three cases of Forestier disease presenting with chief complaint of dysphagia are reported. The reports of all the cases of DISH or Forestier disease presenting with dysphagia, published in English literature are taken for systematic review through WWWS. Information about the Forestier disease is obtained by subject or journal title, title abbreviation, the NML ID and by authors name indexed in PubMed and Google search. We used 'Forestier disease', 'DISH' and 'dysphagia' as key words for network searching. The literature includes full articles and abstracts available on PubMed and Google site.

Case Reports

Case One

A 67 years old male patient, presented with complaint of gradually progressing dysphagia, more to solid for last 1½ year. He lost 12 kg of weight within last 1 year before presentation and on first visit his weight was 59 kg. He had also a complaint of mild pain at the back of neck especially on turning the head side to side and bending the head forward. There was neither any pain during deglutition nor

any voice change. He was hypertensive and had ischemic heart disease, on aspirin, nitrate and ACE inhibitor. On examination he was pale, there was no obvious pathology found in oropharynx. Indirect laryngoscopy revealed no abnormality. There was no palpable node in his neck. Systemic examination was unremarkable. Patient was asked to take some dry food in front of the examiner. There was noticeable discomfort during swallowing and it took an unusual long time to finish the task. Barium swallow X-ray of esophagus revealed a filling defect due to prominent anterior bony spur from fifth, sixth and seventh cervical vertebrae, compressing the hypopharynx and cervical esophagus from the posterior aspect (Fig. 1). The rest of the esophagus was normal. There was no abnormality in routine blood count except 8.7 g% of haemoglobin level. The RBC morphology was hypochromic microcytic. His blood sugar, urea and creatinine levels were within normal limits. Chest X-ray was normal.

As there was no other etiology of dysphagia and the barium swallow X-ray was suggestive, the cause of his symptom was considered to be the Forestier disease affecting the cervical spine. We advised the patient to change the food pattern to semisolid diet with high protein content, to take small and frequent meals and to take plenty of water per mouth. Oral iron supplementation and prokinetic drug (Mosapride) were also advised. After 3 months patient regained 3 kg of weight and faced less discomfort during deglutition while taking semisolid meal. Considering the improvement with conservative measures and the

Fig. 1 Barium swallow X-ray in lateral view showing filling defect due to prominent osteophytes at C5–C7 vertebrae

medical problem of the patient we did not planned any surgical remedy. After 1 year the patient was leading almost normal life with altered food pattern and his body weight was 67 kg.

Case Two

An 82 years old lady was referred to our outpatient department from a district hospital with complaint of pain in neck for 6 years and difficulty in swallowing solid food for last 4 years. An X-ray of cervical spine was done earlier, before development of dysphagia, which was reported as cervical spondylosis according to the patient as the plate was lost. She was treated for that condition by orthopedic surgeon with analgesics and neck exercise. Subsequently she developed dysphagia and was treated with prokinetic drug and proton pump inhibitor for long time with not much improvement. At the time of presentation nutritional status of the patient was poor, her weight was only 42 kg and she was pale and cachectic. Local ENT examination and systemic examination were normal. We performed a flexible esophagoscopy which was normal. On barium swallow X-rays of esophagus there were anterior bony projections from sixth and seventh cervical vertebrae with narrowing of esophageal lumen. Haemoglobin level was 5.9 g% and rest of the blood count was normal. Patient was not diabetic and no other abnormality was found on routine investigations.

We admitted the patient, put her on nasogastric tube feeding and three units whole blood were transfused in 3 days. We used protein supplement powder and other liquids with high nutritional value via the nasogastric tube. After 2 weeks nasogastric tube was withdrawn and the lady was advised to take liquid diet orally and after another week she was advised to take semisolid but high protein food orally. We discharged the patient with advice to take semisolid food in small quantity and in frequent intervals along with tablet mosapride and oral iron. After 3 months her weight was 50 kg with 10.2 g% hemoglobin level and faced considerably less difficulty in taking food as per our advice. We stopped both of her medications at that visit and advised only to continue the modified food habit. After 6 months her weight was 53 kg and was quite happy with her semisolid food as it was not giving her much distress during swallowing.

Case Three

A 75 years old hypertensive male patient from a remote village attended ENT OPD with complaint of lump sensation and foreign body sensation of throat for 2 years. A past history of fish bone impaction in throat was there 2 years back but there was no finding suggesting any

foreign body. He had consulted several physicians and one otolaryngologist for his problem without getting any relief. He was treated with several antibiotics, gargle solutions, acid lowering agents and antidepressants. On careful history taking he said that he also had some difficulty in swallowing as he had to take more time to swallow solid food. Her general built and nutrition was average. No abnormality was found on local or systemic examination. Routine investigations were within normal limits including hemoglobin percentage of 11.9 g%. On plain X-ray of cervical spine there was a large anterior bony projection involving second, third and fourth cervical vertebrae. Barium swallow X-rays revealed slight narrowing of pharyngeal food passage at the same level. Patient was asked to avoid solid food. He was explained about the condition and reassured that the condition was not related to the incidence of fish bone impaction or any other grave disease. Among the medicines he was treated only with prokinetic drug for 3 week. After 1 month patient was almost symptom free. He was advised to continue the soft food. After that the patient has not come for further follow up.

Results and Discussion

Three cases of Forestier disease presenting with dysphagia were identified in our institute from January 2001 to December 2010 and reported. This proves the rarity of the condition. In WWWS only 73 cases were found, published from 1973 to 2010 in different English literatures.

Most of the cases were reported from the Mediterranean region and eastern European countries, mostly from Turkey (16 cases) [2–9] and Italy (13 cases) [10, 11]. Seven cases were reported each from Germany [12–14] and the USA [15–17]. Five cases were reported each from the UK [18–22] and Chile [23]. Eight cases were reported from eastern Asia, four each from Japan and Korea [24–29]. Two case reports were from Netherlands [30]. Singapore, India, France, Austria, Taiwan, Australia, Iran, Saudi Arabia, Puerto Rico and Greece added single case each to the list [31–40]. This geographic distribution pattern is depicted in Fig. 2. This distribution pattern may be due to under recognition of the condition or under reporting.

Among the 73 cases only 11 were female and rest 62 were male (M:F 5.64:1). The disease mainly affects older individuals (29 cases/39.73% in the seventh decade, 23/31.5% in the eighth decade or more, 17 cases/23.29% in the sixth decade and only 4 cases/5.48% were before the sixth decade). Among our cases two were male patients and the other was female. One of our cases was in the seventh decade and rest two were of the eighth decade or above. Our findings regarding age and sex corroborates with the

Fig. 2 Geographic distribution of cases of Forestier disease presenting with dysphagia

result of compiled data. An unique case was reported from Turkey by İlbay et al. [6] where an 11-year-old child was suffering from this condition. It was lowest age incidence found, can be considered as an exception as no other case report were found in young individuals and the next minimum age found in literature was 48 years [12].

In most of the cases, the durations of the problem were considerably long, 42 cases (57.53%) presented after 1 year of initiation of their dysphagia, 10 cases (13.7%) between 6 months to 1 year, 17 cases (23.29%) within 6 months but more than 1 month and only 4 cases (5.48%) presented with short duration of symptom of less than 1 month. All of our patients presented after 1 year of initiation of their symptoms, corroborating with the search result. It signifies that dysphagia due to Forestier disease is commonly progressive in nature and acute presentation is very rare. In the initial phase patient can manage to swallow food with some extra effort and commonly seeks advice from physician when the problem becomes difficult to manage.

Apart from dysphagia which is the commonest presenting problem of the Forestier disease affecting the cervical vertebrae, a number of other problems can also be present. Among the reported cases neck pain is the commonest among them, present in 29 cases (39.73%). Dysphonia was present in 11 cases (15.07%) and respiratory difficulty and stridor due to compression of larynx or trachea in 8 cases (10.96%). The patients often lose their weight due to difficulty in swallowing and reduced food intake. Among the reported cases 8 patients (10.96%) had significant weight loss (>10 kg) within the period between the onset of dysphagia and the time of presentation. Two cases of our series had significant weight loss and one of

them was cachectic. Another potentially dangerous complication, aspiration pneumonia was present in 4 (5.48%) patients among the 73 reported cases. Fortunately none of our patients had this complication. Some other non-specific symptoms like foreign body sensation in throat, lump sensation in throat and reduced neck mobility were also present in some of the reported cases. Two case reports needs special mention in this context of associated symptoms. One was reported by Tomoyuki et al. [24], the patient was presented with a neck tumor similar to advanced thyroid carcinoma on palpation and on ultrasonography. Another case experienced a syncopal attack followed by quadriplegia, reported by Gun Seok et al. [28].

Most of the reported cases were investigated with plain X-ray of the neck in lateral view (57 cases/78.09%) and barium swallow X-ray (50 cases/68.49%). CT scan and MRI were done in 29 (39.73%) and 16 cases (21.92%), respectively. Upper G.I endoscopy was done in 26 cases (35.62%) and 8 patients (10.96%) were investigated with video fluoroscopy. In our series, barium swallow X-ray was done in all the three patients, which concluded the diagnosis. In one patient plain neck X-ray was done as a primary step and in another case flexible esophagoscopy was done.

As per gathered report from literature, the condition mainly affects the lower cervical vertebrae, especially C3–C6 region and often involves multiple vertebrae. Maximum no. of cases involved C4 and C5 vertebrae. Gradually lesser number of cases affected both upper and lower level vertebrae (Fig. 3). An exceptional case reported by Underberg-Davis and Levine [17], where a giant osteophyte at T9–T10 level caused acute onset dysphagia in a 76 years old man.

Fig. 3 Line diagram depicting no. of cases affecting different level of vertebrae

Most of the reported cases were treated by surgical correction of the causative factor (53 cases/72.6%). Antero-lateral neck approach was considered by most of the authors. 46 cases (86.79%) were relieved of the problem with surgical treatment. As the disease affects mainly the older individuals, there are often some surgical contraindicating factors like very old age, heart disease, and diabetes mellitus etc. which limit the scope of surgery. Those cases were treated with conservative approaches like swallow training including encouraging semisolid diet and taking more time for eating. Outcome of these cases were favourable as 14 cases out of 17 (82.35%) managed to maintain their nutritional status. All three cases of our series were with contraindications for surgery. Our approach was conservative with asking the patients to change their food habit towards soft diet, taking plenty of water with food, taking frequent meal with small quantity at a time and taking time to complete the meal. All three patients improved with these measures. Two of the cases reported in literature undergone palliative surgery in form of feeding gastrostomy [18, 29]. One of them died of some unrelated condition [18]. A new modality of treatment for dysphagia due to cervical osteophytes was reported by Unlu et al. [8] in 2008. They used phonophoresis (local application of NSAID gel along with ultrasonic ray locally) and achieved significant improvement in seven out of nine of their patients.

Conclusion

DISH is an uncommon cause of dysphagia mostly affects older individuals and with male preponderance. The condition is reported maximally from the countries around the Mediterranean Sea till date. Lower cervical vertebrae are mostly affected. The disease usually presented with a long

course, often with some other associated symptoms like neck pain, reduced neck mobility, voice change, stridor or even aspiration related complications. Main and definitive treatment modality is surgical removal of the osteophytes. If surgery is contraindicated, change of food habit and swallow training can be advised and can be of value in maintaining nutritional requirement. Our series corroborates the consensus about the patient profile and the level of vertebrae involved. But none of our patients was treated surgically as their general condition or medical problems did not permit, but change of food habit and swallow training was fruitful in all of the cases.

Conflict of interest The authors declare presence of no conflict of interest.

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