Pharyngoesophageal junction neoplasia - therapeutic management

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Abstract
Not only the anatomy but the treatment and the prognosis of these tumors are intermediate between hypopharyngeal and esophageal tumors. After a portion of the esophagus is removed or complete esophagectomy, a conduit must be established. The authors reviewed the experience of Prof. Cristian Popescu in total pharyngo-laryngectomy and his technique of pharyngoesophageal reconstruction with synthetic esophageal prosthesis. We have some 21 patients who underwent an esophageal reconstruction with Montgomery esophageal tube. This is a very important new, modern, interdisciplinary approach between the head and neck surgeon and the general / thoracic surgeon to treat the pharyngo-laryngo-esophageal neoplasia with one stage reconstruction. The follow up for these patients shows that this reconstruction method is a good, reliable choice with low costs and considerable advantages for the quality of life. Surgery for these patients should be considered primarily palliative and the optimal reconstruction should preserve the quality of life for the duration of survival.

Key words: pharyngo-esophageal region, neoplasia, reconstruction, synthetic prosthesis

Introduction
The pharyngo-esophageal junction, also called “the mouth of the esophagus” represents the passage from the hypopharynx to the cervical esophagus. The anatomy, treatment and prognosis of this region are intermediate between the hypopharyngeal and esophageal neoplasia (1). The diagnostic of this type of neo-
plasia is variable in regards to the circumstances of apparition and the neoplastic growth in this region (2).

Lying at about 15-16 cm from the dental arch in occlusion the mouth of the esophagus is lined anteriorly by the seal of the cricoid cartilage, inferiorly by a plane that underlies the inferior margin of the body of the cricoid cartilage and superiorly by the insertion of the cricoaritenoid and interaritenoid muscles (they belong to the hypopharynx). This section passes thorough the middle third of the seal of the cricoid cartilage. Posteriorly we intersect a crest of pharingo-esophageal mucosa and in the lateral region lies the inferior pharangeal constrictor muscle (the crico-pharingeal region). The cricoid gorge of the esophagus is due to the disposition of the muscular fibers of the inferior pharangeal constrictor muscle (pars fundiformis) (6).

The indirect laryngoscopy is useless in examining this passage postcricoid region towards the cervical esophagus although Killian described a method for examining the mouth of the esophagus (Fig. 1)(3).

Rigid endoscopy is the elected method of examination to asses information on this region, the mouth of the esophagus beeing the first region of the esophagus that presents resistance to the forward movement of the endoscope (between the cricoid seal and the posterior pharingeal wall). Perforative accidents of the posterior wall of the pharynx are likely to occur in the pharingo-esophageal junction (Fig. 2,3)(4).

The risk factors for the junction neoplasia are the same as in the hypopharangeal neoplasia: alcohol (80%), tobacco (65%), food habits, Plummer-Vinson syndrome, heredocolateral antecedents. It has been noticed that a low age of apparition of hypopharangeal and cervical esophagus neoplasia in patients with family history of neoplasia. There is no clear link between lifestyle and hypopharangeal and cervical esophagus neoplasia. Malignant degeneration is found in pharingoesophageal diverticulum, corrosive esophagitis, Patterson-Kelly syndrome pathological conditions. The mouth of the esophagus can be invaded by a desendent growing hypopharangeal cancer (piriform sinus, postcricoid region, lateral or posterior walls), seldom by local extension (cricoid chondrosarcoma) or by ascending cervical esophagus cancer. Primitive neoplasia that are localized at the pharingoesophageal junction are seldom fast diagnosed because the mouth of the esophagus is hard to visualise while attempting indirect laryngoscopy; We have to take into consideration the multiple neoplasia syndrome because the junction neoplasia is frequently associated with other level neoplasia (16).

From the standpoint of histopathology pharingoesophageal neoplasia are usually spinocellular carcinomas. In the E.N.T. Department of Coltea Clinical Hospital we preponderantly found differentiated forms of carcinoma some 54% beeing well differentiated infiltrant spinocellular carcinomas, 28% moderate differentiated carcinomas and the rest of 18% beeing poorly differentiated carcinomas.

The tumors of the postcricoid regions invade the postreior wall of the pharynx and from time to time precociously the apex of the piriform sinus. The lymphatic spread is usually made via jugulocarothid ganglia and secondarily via spinal ganglia. (5)
The cervical esophagus tumors spread directly, lymphatically and by blood stream. Prof. Dan Gavriliu claimed that the gaps of the lymphatic network from the mucosa and the muscular layers in the esophagus are oblonged which explains why malignant tumors have the tendency of spreading tumoral cells intramuraly away from the original macroscopic neoplastic site (7). This fact explains the use of total pharyngolaryngoesophagectomy in this type of tumors. According to Cordos the macroscopic caudal extension of the tumor is no more than 4 cm (17).

It is considered that T1 junction cancer does not exist, the staging beginning with T2 stage no matter the size of the tumor. Generally tumors are being diagnosed in T3-T4 stages. According to Mouhnier-Khun T3 stage stands for limited anterior wall lesion but with anterograde and retrograde extension, or a mouth limited lesion but with circular extension. T4 stage is mentioned as a circular lesion that extends beyond the junction or a nearby structures invading lesion (Fig. 4,5,6,7,8) (16).

The symptomatology of esophageal tumors is governed by dysphagia selective in the beginning for solid food and total afterwards depending on the evolution, thus leading to the alteration of the nutritional status. The general status is good at the beginning of the illness. The patient complains about pharyngeal paresthesia, foreign body sensation, fetid halitosis, regurgitations and eructation. The association between dysphagia with “false way” sensation is reminiscent for neoplasia of the mouth of the esophagus. Superior digestive bleeding appear in ulcerated tumors. Dysphonia (recurrent paralysis), dyspnoea (sublaryngeal compression of the posterior tracheal wall), Claude-Bernard-Horner syndrome, prelarynx ganglia (Poirier ganglion), jugulo-carotid ganglia and supraclavicular ganglia (Troisier ganglion on the left side), the disparition of the laringeal cracment are all signs and symptoms noticeable while examining the cervical region. There is also the possibility of developing liver, lung, brain and bone metastases in the sickness evolution.

The paraclinical diagnosis is made using standard and contrast substance Rx scans, CT scan, MRI. The endoscopic investigation (suspended laryngoscopy, esophagoscopy and bronchoscopy) with biopic material prelevation and luminal ultrasound examination are essential for diagnostic and tumoral staging. The diagnostic certainty is made after histopathological and immunohistochemical analysis (Fig. 9)(4).

Materials and Methods

The treatment for the pharyngo-esophageal junction neoplasia is complex and multimodal that requires the participation of the E.N.T. surgeon, radiotherapist, chemotherapist, nutritionist and psychologist. From a surgical standpoint there are two major problems to be taken into consideration: excision and pharyngosophageal reconstruction. While excising the esophagus partially or totally the preservation of the larynx is virtually impossible. Thus the need for performing a total circular pharyngolaryngectomy with partial or total eso-phagectomy depending on the tumoral extension with bilateral neck dissection (12,13), followed by per primam reconstruction of the digestive tract (8,9).

Partial pharyngo-esophagectomies (posterior and posterolateral) are only indicated in T2 stage tumors or primitive junction tumors followed by reconstructive techniques using the larynx (Labayle or Som techniques).
It is considered that a circumferential defect is a 70% loss in the pharynx circumference or when postexcision pharyngeal mucosa is less than 2 cm. In these situations there have been described a series of reconstructive techniques using free skin grafts (Conley technique), reversed cutaneous cervical flaps (Andre-Laccourreye, Lewis, Bertain technique), thoracic cutaneous flaps (Pietrantoni, Bakamjian), musculo-cutaneous tubulised flaps out of great pectoral muscle, free tubulised radial flap, free transfer of jejunal segment (8,9). In case of total esophageal resection the reconstruction is made by gastric pull-up or bowel transposition (11).

From 2000 in the E.N.T. Department of Coltea Clinical Hospital Prof. C.R. Popescu rehabilitates the superior digestive tract, after circular pharyngo-laryngectomies, by using Montgomery prosthesis (21 cases). Before the use of such prosthesis the rehabilitation was tempted by using prosthesis made out from different types of materials (dacron, plastic) but were rejected by the human organism. Pharyngo-esophageal prosthesis are made out of silicone with different sizes and radiotransparency degrees. In the tubular segment the prosthesis has two dilated regions that offer better bed stabilisation and the drainage of saliva no matter the position of the head. We use Montgomery prosthesis for better quality of life and intermediate step to free flap reconstruction. (10)

At the cranial end the prosthesis has a funnel shape that is sutured at the root of the tongue and the oro-/hypopharynx. The caudal end of the prosthesis is placed directly into the lumen of the esophagus (Fig. 10,11) (14,15).

For prosthesis hothead stabilisation it is very important that we preserve and mend the prelarynx muscles. X-ray analysis allows the check-up for prosthesis positioning and permeability (Fig. 12,13).

Postreconstruction control can also be made by endoscopy visualising the cranial end of the prosthesis sutured at the root of the tongue and the caudal end in the esophagus (Fig. 14,15).

Because of a missized prosthesis there can occur decubitus lesions on the esophageal mucosa at the inferior end with the forming of a granulation tissue that impairs normal oral feeding. This lead to the surgical reintervention for replacing the prosthesis with a smaller one (Fig. 16).

Histopathological examination of the tissue formed at the inferior end of the prosthesis ruled out the possibility of tumoral relapse. Another complication for this reconstructive method is the appearance of a fistula between the pharynx and the tegument especially at the cranial end of the prosthesis because precocious oral feeding of low compliant patients (less than 10 to 14 days postreconstruction).

We encountered a series of complications post-prosthesis reconstruction:
- pharyngo-cutaneous fistulae - 3 cases (anterior, at the root of the tongue, lateral) (Fig. 17-18);
- dissolution of sutures at the cranial end of the prosthesis - 1 case (Fig. 19);
- psychological intolerance with suicidal attempt – 1 case.

Radiotherapy tolerance is comparable to that after other...
reconstructive techniques. Radiotherapy doses administered to patients that underwent such a reconstructive technique were an average of 65 Gy in comparison to 40 Gy used after gastric pull-up, 70 Gy used after musculo-cutaneous flaps reconstruction and 60 GY used after bowel transposition.

The mean hospitalisation period was 2 to 3 weeks and the oral feeding can be restored after 10 to 14 days. The nasogastric tube fitted intraoperatory thorough the Montgomery prosthesis lumen was suppressed after performing the methilen blue administration.
There were no intraoperatory deaths and the postoperative mortality is high by local tumoral relaps, the invasion of the jugulo-carotid vessels with massive bleeding (1 case) and metastatic spread (pulmonary metastases - 1 case).

In conclusion, the advantages of using this type of prosthesis are:
- reduced time of the surgery procedure;
- increase in quality of life;
- reduced hospitalisation costs;
- performed in one step;
- low cost;
- reduced mortality;
- reduced mobility;
- post-op radiotherapy tolerance.

High grade relapsing tumors have a certain indication for associated concurrent radiotherapy. There are no indications of lifespan prolongation for preoperative radio- (20-40Gy) or chemotherapy.

Clinical nutrition is a must in case of such patients in regards to their nutritional status evaluated at admission into our department using the BMI and the NRS 2002 protocol. Clinical nutrition must be continued throughout the entire period of hospitalisation while evaluating clinical parameters for each patient (required caloric intake, biochemistry constants). Enteral nutrition via naso-gastric tube jejunal or gastric stoma with standardized nutritional supplements according to ESPEN 2006 guidelines needs to be initiated as soon as possible if the digestive tract is functional (18).

Conclusions

Despite modern methods and techniques of diagnose and staging, different types of surgical excision and reconstructive techniques, development in the radiotherapy and chemotherapy protocols there is still a poor prognostic for hypopharangeal and cervical esophagus cancer. One of the most important modern concerns in the management of this type of neoplasia is the multimodal therapy that includes different medical and surgical specialities, including the psychologist. Surgical efforts can be considered to be paliative from the very beginning considering survival rates at 3 and 5 years therefore our main goal is to improve the quality of life for these oncological patients. The best medical approach is dictated by different sets of factors that include general status of the patient, local and systemic implications, type of neoplasia, medical resources and last but not least the acceptance of illness by the patient and his approval for medical attention. Different reconstructive techniques are available and futher improvement of biomaterials lead to advance and new surgical techniques.

References