Gastric adenocarcinoma of the upper oesophagus: A literature review and case report

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A B S T R A C T
BACKGROUND: Ectopic gastric mucosa (EGM) otherwise termed gastric heterotopia or gastric inlet patch occurs in approximately 2.5% of the population. Adenocarcinoma uncommonly involves the upper oesophagus, rarely arising from gastric heterotopia or submucosal glands. Currently, there are 58 cases in the literature of oesophageal adenocarcinoma arising within areas of EGM. To date no paper has differentiated between gastric or intestinal type adenocarcinoma. This case, which describes adenocarcinoma arising within EGM, exhibited a different immunophenotype reminiscent of gastric type glands, in the absence of intestinal metaplasia. This case should be regarded as a different type of carcinoma, consistent with a non–Barrett’s oesophagus-associated adenocarcinoma.

CLINICAL PRESENTATION: A 63 year old female presented with a three month history of progressive cervical dysphagia with no associated weight loss or general malaise. Gastroscopy revealed a suspicious lesion at the cricopharyngeus. Positron emission tomography demonstrated a metabolically active primary lesion without evidence of distant disease. The patient received neo-adjuvant chemotherapy followed by a three stage total oesophagectomy. Histology demonstrated a moderately differentiated adenocarcinoma with gastric immunophenotype and background changes of gastric heterotopia.

CONCLUSION: EGM is common but scarcely biopsied for evidence of dysplasia or adenocarcinoma. Whilst malignant progression is rare it is important that endoscopists are aware of the potential. Determining the exact type of adenocarcinoma may have implications for therapeutic approaches. Recognition of EGM at endoscopy may identify patients at greater risk of developing adenocarcinomas of the proximal oesophagus; however, this relationship and the necessity for screening requires more study.

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1. Introduction

The incidence of oesophageal cancer is increasing and currently affects in excess of 450,000 people each year worldwide [1]. Most commonly squamous cell carcinoma arises within the proximal third of the oesophagus and less often adenocarcinoma occurs within the distal third of the oesophagus in association reflux and Barrett’s metaplasia. Adenocarcinoma within the proximal oesophagus and unrelated to Barrett’s metaplasia are extremely rare and arise either from foci of EGM or submucosal glands. At least 58 cases of oesophageal carcinoma arising in an area of ectopic gastric mucosa (EGM), also referred to as gastric inlet patch or gastric heterotopia, have been reported in the literature. However this is the first to describe an adenocarcinoma arising within the proximal oesophagus within an area of EGM that has exhibited gastric immunophenotype.

1.1. Pathogenesis of ectopic gastric mucosa

The aetiology of EGM is poorly understood. Currently, two main theories exist to explain the pathogenesis. The most widely accepted of these is that EGM is an embryological remnant. Incomplete embryological replacement of columnar mucosa by squamous epithelium leads to the development of remnant patches of columnar epithelium which differentiate into gastric mucosa [2,3]. The alternative theory proposes that EGM, in a similar way to Barrett’s oesophagus, is an acquired condition as a result of gastro-oesophageal reflux disease [4]. One study found that the immunohistochemical staining of an adenocarcinoma arising

Abbreviations: EGM, ectopic gastric mucosa; CK, cytokeratin; PET, positron emission tomography; FDG, fluorodeoxyglucose; CT, computed tomography; EC, Epirubicin, cisplatin, fluorouracil 5FU; MUC, mucins.
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within a patch of EGM, shared the same pattern of staining as that of Barrett’s metaplasia, that is CK7 positive and CK20 negative staining. They concluded from this that EGM and Barrett’s must have a common pathogenesis. However, whilst EGM itself may be a congenital condition, exposure to acid, either secreted from parietal cells within the EGM or refluxing caudally from the stomach, may lead to the acquired changes known to cause Barrett’s and hence explain the immunohistochemical staining.

1.2. Development of invasive carcinoma within areas of ectopic gastric mucosa

It is widely accepted that Barrett’s oesophagus can lead to the development of adenocarcinoma as a result of successive dysplastic changes culminating in invasive carcinoma [5]. This metaplasia-dysplasia sequence could also be the cause of adenocarcinomas arising in EGM, especially considering that EGM likely contains parietal cells that secrete hydrochloric acid locally, and thus induce the metaplasia-dysplasia pathway. However, in this case, no intestinal metaplasia was found histologically and the presence of EGM raised the possibility of a different pathway of carcinogenesis in this tumour.

2. Presentation of case

A 63 year old female initially presented with a 4 month history of progressive cervical dysphagia with no associated weight loss or malaise. Risk factors for cancer included being an ex-smoker and daily alcohol consumption. Her other medical history included hypertension and hypercholesterolaemia.

2.1. Investigations

- Barium swallow – demonstrated a smooth lesion at the level of the cricopharyngeus extending from the inferior border of C6 to inferior border of T2. The remaining oesophagus appeared normal.
- Gastroscopy – confirmed the presence of a suspicious lesion within the upper oesophagus situated immediately below the cricopharyngeus as well as mild oesophagitis, prepyloric gastritis and duodenitis. H. pylori and standard biopsies were obtained.
- Histopathology – a moderately differentiated adenocarcinoma of gastric type with variable glandular, villiform and papillary architecture with marked cytological and nuclear atypia, as well as marked inflammation was found in the biopsies taken from the proximal oesophageal lesion.
- Staging CT – invasion into the adjacent mediastinal fat but no definite invasion of the great vessels or trachea and no definite lymphadenopathy could be identified.
- PET scan – revealed an FDG-avid right sided para-oesophageal lymph node and confirmed the absence of any distant FDG-avid disease.
- Bronchoscopy and endobronchial ultrasound were also performed and demonstrated no invasion of the airway structures.
- Clinical staging was cT3 N0.

The patient was discussed at a multidisciplinary meeting and the decision was made to proceed with neoadjuvant chemotherapy. The patient received ECF neoadjuvant chemotherapy.

2.2. Operative findings and technique

After completion of induction chemotherapy the patient underwent a three stage total oesophagectomy.

A 2 cm tumour within the upper oesophagus was found 19 cm distal to the oral cavity and 5 cm distal to the pharynx. There was no evidence of macroscopic metastatic disease in the abdomen, neck or chest. Enlarged, but soft, para-oesophageal nodes where noted in the upper mediastinum.

Thoracoscopic mobilisation of the oesophagus was performed with clearance of the upper para-oesophageal nodes. A left neck incision was made along the anterior border of sternocleidomastoïd through which the oesophagus was completely mobilised up to the larynx. At this point, it was felt that the tumour could be completely resected without performing laryngectomy. A prophylactic tracheostomy was performed.

The cervical oesophagus was divided 2 cm proximal to the tumour. A single layered end to side, oesophago-gastric anastomosis was fashioned with 3.0 PDS. A nasogastric tube was placed across the anastomosis.

A pyloromyotomy was performed and a feeding jejunostomy placed.

2.3. Histology

Histological examination of the specimen revealed a non-Barrett’s associated gastric type adenocarcinoma arising within an area of ectopic gastric mucosa in the upper oesophagus (Fig. 1), in the absence of any intestinal metaplasia. Adjacent areas of high grade dysplasia of gastric/foveal type were noted.

Immunohistochemical staining revealed the tumour was CK7 strongly and diffusely positive (Fig. 2), with only patchy-weak CK20 expression. Also, there was strong diffuse expression of MUC-1 (Fig. 3), as well as MUC-SAC (Fig. 4) but only sparse MUC-2 staining and no CDX-2 labeling. This immunoprofile and lack of Barrett’s oesophagus is consistent with non-intestinal phenotype adenocarcinoma of the upper gastro-intestinal tract. The tumour was limited to the deep submucosa with no invasion into the muscularis propria or lymph node involvement. Pathological stage was ypT1b N0.

3. Follow up

The patient is currently disease free 20 months post surgery. The patient had gastroscopies 2–5 months post operatively for management of dysphagia secondary to anastomotic sticture. These procedures demonstrated no evidence of macroscopic tumour recurrence. Notably, no were biopsies obtained. The last surveillance CT scan demonstrated no evidence of disease recurrence. 11 months post surgery the patient presented with nausea and vomiting secondary to small bowel obstruction. An emergency laparotomy and adhesiolysis was performed which also confirmed the absence of any intra-abdominal metastasis.

4. Literature review

We searched Ovid and Embase databases using the search terms ‘ectopic gastric mucosa’, ‘heterotopic gastric mucosa’, gastric inlet patch’ and ‘adenocarcinoma’. We also yielded results from the reference lists of papers identified in this search.

5. Results

In total 58 cases of adenocarcinoma arising within EGM in the proximal oesophagus were identified in 52 papers (Table 1). Two papers could not be obtained in English full text, Frezza et al. and Armstrong et al. In the majority of papers the finding of intestinal metaplasia was not reported on (n = 42). Only 3 cases reported finding intestinal metaplasia and 7 cases reported on the absence of intestinal metaplasia. Cases of adenocarcinoma arising EGM have been reported worldwide (Table 2), interestingly the majority of cases have been reported in Japan. Median age 64 (43–88) years
Table 1  
Table of all papers reporting cases of adenocarcinoma arising within ectopic gastric mucosa.

| No. | Paper                              | Country | Year | Age | Gender | TNM         | LVI  | Histology                          | Intestinal Metaplasia | Treatment                          | Follow Up                        |
|-----|------------------------------------|---------|------|-----|--------|-------------|------|------------------------------------|-----------------------|------------------------------------|-----------------------------------|
| 1   | A Carrie (Carrie:1950vk)           | UK      | 1950 | 64  | M      | pT2NXM0     |      | Adenocarcinoma                     | ?                     | Subtotal oesophagectomy            | 1 year, no recurrence, alive      |
| 2   | BC Morson (Morson:1952ci)          | UK      | 1952 | 56  | M      | cT3N1M1     |      | Adenocarcinoma (high grade)        | N                     | Subtotal oesophagectomy            | 2 months, alive                   |
| 3   | Frezza LG                          | Italy   | 1956 |     |        |             |      |                                    |                       |                                    |                                   |
| 4   | Armstrong                          |         | 1959 |     |        |             |      |                                    |                       |                                    |                                   |
| 5   | H A Raphael (Raphael:1966ci)      | USA     | 1966 | 75  | M      |             |      | Adenocarcinoma (grade 3)           |                       | Radiotherapy and dilatation        | 34 months, died                   |
| 6   |                                    |         |      |     | F      |             |      |                                    |                       |                                    |                                   |
| 7   |                                    |         |      |     |        |             |      |                                    |                       |                                    |                                   |
| 8   | W M Davis (Davis:1969ts)          | USA     | 1969 | 68  | M      | pT1NXM0     |      | Adenocarcinoma, mucinous           |                       | Radiotherapy and dilatation        | 7 months, alive, no recurrence    |
| 9   | G Sakamoto (Sakamoto:1970uo)      | Japan   | 1970 | 64  | M      | pT2N0M0     |      | Adenocarcinoma                     |                       | Oesophagectomy                     | 10 months, died                   |
| 10  | P Jernstrom (Jernstrom:1970vs)    | USA     | 1970 | 73  | M      | pT3N0M0     |      | Poorly differentiated adenocarcinoma|                       | Radiotherapy and oesophagectomy   | 4 months, died                    |
| 11  | C Clemente (Clemente:1974up)      | Italy   | 1974 | 53  | M      | pT3         |      | Adenocarcinoma                     |                       | Oesophagectomy                     | 10 months, recurrence             |
| 12  | B Danoff (Danoff:1978vk)          | USA     | 1978 | 43  | M      | cT4N0M0     |      | Poorly differentiated adenocarcinoma|                       | Radiotherapy                       | 9 months, died                    |
| 13  | H Schmidt (Schmidt:1985ts)        | Germany | 1985 | 54  | M      |             |      |                                    |                       |                                    |                                   |
| 14  |                                    |         |      |     |        |             |      |                                    |                       |                                    |                                   |
| 15  | O Goenau-Brissonniere (GoenouBrissonniere:1985wi) | France | 1985 | 38  | M      | pT3         |      | Adenocarcinoma                     |                       | Oesophagectomy                     | 4 months, died                    |
| 16  | W N Christensen (Christensen:1987wi) | USA    | 1987 | 52  | M      | pT2N1M0     |      | Poorly differentiated adenocarcinoma|                       | Oesophagectomy                     | 25 months, recurrence             |
| 17  |                                    |         |      |     |        |             |      |                                    |                       |                                    |                                   |
| 18  |                                    |         |      |     |        |             |      |                                    |                       |                                    |                                   |
| 19  | K Ishii (Ishii:1991tn)            | Japan   | 1991 | 66  | M      | cT3N7       |      | Well differentiated tubular adenocarcinoma | N | Oesophagectomy                     | 20 months, alive no recurrence    |
| 20  | Takagi Y (Takagi:1994gc)          | Japan   | 1994 | 85  | M      | A2NXM0Ptx   |      | Poorly differentiated adenocarcinoma |                       | Radiotherapy                       | Unknown                           |
| 21  | Kubota S (S:1993tc)              |         | 1993 | 58  | M      |             |      |                                    |                       |                                    |                                   |
| 22  | R M Sperling (Sperling:1995uf)    | USA     | 1995 | 79  | M      | cT4N0M0     |      | Poorly differentiated adenocarcinoma|                       | Radiotherapy                       | Unknown                           |
| No. | Paper                                 | Country          | Year | Age | Gender | TNM    | LVI | Histology                          | Intestinal Metaplasia | Treatment                           | Follow Up              |
|-----|---------------------------------------|------------------|------|-----|--------|--------|-----|------------------------------------|------------------------|-------------------------------------|------------------------|
| 23  | Takagi et al. (Takagi:1995tw)          | Japan            | 1995 | 70  | M      | pT1N0M0 | N   | Well differentiated tubular adenocarcinoma | ?                      | Oesophagectomy                      | Unknown                |
| 24  | Mion F et al. (Mion:1996et)            | France           | 1996 | 45  | M      | uT1N0MX | N   | Tubulovillous adenoma              | with high grade dysplasia | Subtotal oesophagectomy             | Unknown                |
| 25  | Kamitori et al. (Takagi:1995tw)        | Japan            | 1996 | 74  | F      | pT1N0M0 | N   | Well differentiated adenocarcinoma  |                        | Subtotal oesophagectomy             | Unknown                |
| 26  | S Pai et al. (Pai:1997wc)              | India            | 1997 | 60  | M      | pT2N0M0 | N   | Poorly differentiated adenocarcinoma |                        | Oesophagectomy and chemoradiotherapy | 24 months, recurrence |
| 27  | C Berkelhammer et al. (Berkelhammer:1997ik) | USA             | 1997 | 71  | M      | pT1bN1M0 | N   | Moderately differentiated adenocarcinoma |                        | Oesophagectomy                      | 24 months, alive, no recurrence |
| 28  | G V Lauwers et al. (Lauwers:1998us)   | USA              | 1998 | 57  | M      | cT4or3, N0,M0 | N   | Moderately differentiated adenocarcinoma, mucinous and papillary | Y                     | Oesophagectomy, total laryngectomy and partial pharyngectomy, adjuvant radiotherapy | 8 months, alive, no recurrence |
| 29  | O Pech et al. (Pech:2001wo)            | Germany          | 2001 | 77  | M      | pT1bN0M0 | N   | Well differentiated adenocarcinoma  |                        | EMR                                | 12 months, alive, no recurrence |
| 30  | J M Klaasen et al. (Klaasen:2001ki)    | Netherlands      | 2001 | 66  | M      | pT4N1M0 | N   | High grade dysplasia               | within EGM              | Endoscopic mucosal ablation         | 16 months, alive       |
| 31  |                                                | Spain            | 2001 | 43  |        | pT4N1M0 | N   | Poorly differentiated adenocarcinoma |                        | Subtotal oesophagectomy             | 4 months, died          |
| 32  | T Noguchi et al. (Noguchi:2001ww)      | Japan            | 2001 | 73  | M      | pT1NXMX | ?   | Well differentiated adenocarcinoma  |                        | Subtotal oesophagectomy, laryngectomy and pharyngectomy | 5 years, no recurrence |
| 33  | Denis et al. (Chatelain:2002wp)         | France           | 2002 | 61  | M      | cT4N0M0 | ?   | Poorly differentiated adenocarcinoma |                        | Subtotal oesophagectomy             | 15 months, died, no recurrence |
| 34  | Jean-Michel et al. (Balamon:2003wn)    | France           | 2003 | 61  | M      | pT4N0M0 | N   | Adenocarcinoma                      |                        | Oesophagectomy                      | 21 months, died       |
| 35  | Nobuo et al. (Hirayama:2003hy)         | Japan            | 2003 | 77  | M      | pT1N0M0 | N   | Well differentiated papillotubular adenocarcinoma |                        | EMR                                | 31 months, alive       |
| 36  | Tatsuya Abe et al. (Abe:2004ue)        | Japan            | 2004 | 50  | M      | pT1bN0M0 | N   | Well differentiated adenocarcinoma  |                        | Oesophagectomy                      | 18 months, alive, no recurrence |
| 37  | Kagawa et al. (Kagawa:2004un)          | Japan            | 2004 | 51  | M      | ?      | ?    | Moderately differentiated adenocarcinoma |                        | Subtotal oesophagectomy             | 11 months, died, recurrence |
| 38  | S J Afrawi et al. (Afrawi:2005vz)      | USA              | 2005 | 60  | M      | pT1N0M0 | N   | Moderately differentiated adenocarcinoma |                        | Subtotal oesophagectomy, adjuvant chemoradiation | 6 years, alive       |
| 39  | von Rahden et al. (vonRahden:2005cw)   | Germany          | 2005 | 52  | M      | cT3N1M0 | N   | Moderately differentiated tubulo-papillary adenocarcinoma |                        | Oesophagectomy                       | 3 years, alive         |
| 40  | Hayashi et al. (Hayashi:2005st)        | Japan            | 2005 | 72  | F      | cT1N0M0 | N   | Low grade dysplasia in an adenoma, arising in EGM |                        | EMR                                | 3 years, alive          |
| 41  | Hakan Alagozlu et al. (Alagozlu:2007ku) | Turkey         | 2007 | 57  | M      | cT4N1M0 | N   | Poorly differentiated adenocarcinoma |                        | Oesophagectomy                      | Lost to FU             |
| 42  | Hoshino et al. (Hoshino:2007ef)        | Japan            | 2007 | 74  | M      | pT3N0M0 | N   | Adenocarcinoma, papillary          |                        | Oesophagectomy                      | 5 months, recurrence   |
| Case ID | Name            | Country   | Year | Age | Gender | Stage | Histology                               | Treatment                  | Follow-up            |
|---------|-----------------|-----------|------|-----|--------|-------|-----------------------------------------|----------------------------|----------------------|
| 43      | Shuji Komori    | Japan     | 2010 | 75  | M      | pT2N0M0 N | Moderately differentiated tubular adenocarcinoma | Subtotal oesophagectomy       | 3.5 years alive, no recurrence |
| 44      | Yoshida T       | Japan     | 2010 | 79  | M      | cT1N0M0, N | Moderately differentiated adenocarcinoma      | EMR                        | 2 years, alive, no recurrence |
| 45      | Hirano A        | Japan     | 2010 | 57  | M      | ?        | Adenocarcinoma                           | Subtotal oesophagectomy       |                      |
| 46      | Ando T          | Japan     | 2010 | 64  | M      | pT1aN0M0 | Well differentiated adenocarcinoma          | Subtotal oesophagectomy       |                      |
| 47      | Bard A          | France    | 2011 | 87  | M      | cT3N1M0  | Moderately differentiated adenocarcinoma    | EMR                        | 17 months, died      |
| 48      | Hitaka D        | Japan     | 2011 | 64  | M      | uT1N0M0, pT1aN0M0 | Well differentiated adenocarcinoma | Subtotal oesophagectomy       |                      |
| 49      | Toshihiro Kitajima | Japan     | 2011 | 64  | M      | pT1bN0M0 N | Well differentiated tubular adenocarcinoma, mixed neuroendocrine tumour | Radical oesophagectomy       | 16 months, alive      |
| 50      | K Nonaka        | Japan     | 2013 | 74  | M      | pT1aN0M0 | Asymptomatic glandular structures arising in EGM | EMR                        | Unknown              |
| 51      | Yash P Verma    | India     | 2013 | 50  | F      | ?        | Poorly differentiated adenocarcinoma        | Radiotherapy                | 3 months, alive      |
| 52      | Naoki Akanuma   | Japan     | 2013 | 57  | M      | pT2N0M0 N | Well – moderately differentiated adenocarcinoma, positive margins | Neoadjuvant chemo-rad Oesophagectomy Adjuvant chemo-rad | 4 years, alive, no recurrence |
| 53      | Oliver Moschler  | Germany   | 2014 | 83  | M      | pT1aN1M0 Y | Well differentiated adenocarcinoma          | EMR                        | Unknown              |
| 54      | Mariko Tanaka   | Japan     | 2014 | 76  | M      | pT1N0M0 N | Well differentiated tubular adenocarcinoma   | Subtotal oesophagectomy       | 19 months, alive      |
| 55      | Bulent Yasar    | Turkey    | 2014 | 52  | F      | pT1N0M0 N | Well differentiated adenocarcinoma          | EMR                        | 3 months, alive      |
| 56      | V R Hudspeth    | USA       | 2014 | 77  | M      | cT1N0M0  | Moderately differentiated adenocarcinoma    | EMR and argon plasma coagulation to margins RFA | 12 months, alive, no recurrence |
| 57      | Payne J         | USA       | 2015 | 88  | M      | cT7N2M1  | Moderately differentiated adenocarcinoma    | Palliative Chemo-rad        | 10 months, alive      |
| 58      | Amjal S         | USA       | 2015 | 57  | M      | pT1bN0M0 ? | Well – moderately differentiated adenocarcinoma | Oesophagectomy               | Unknown              |
6. Discussion

Ectopic gastric mucosa is a well-recognised phenomenon, however, adenocarcinoma of the proximal oesophagus remains uncommon and many specialist upper gastrointestinal surgeons will never come across a case. The cases summarised here serve as an important reminder to endoscopists to be aware of gastric inlet patches, whose prevalence has been estimated between 0.21% and 10% of the general population[7–9]. The prevalence of cancers arising within EGM does not warrant routine surveillance but attention should be paid to any visualised mucosal abnormality and in selected cases the use of narrowband imaging and specialized chromoendographic stains may be useful. Adenocarcinomas arising within EGM in the cervical oesophagus can often be missed by swiftly passing the scope through the upper oesophagus. Published literature demonstrates that, when found early, it is possible to successfully manage these cancers with endomucosal resection. In particular situations, for example where patients require prophylactic gastrectomy for genetic predisposition to gastric cancer (e.g. CDH-1), knowledge of EGM may prove important.

(Table 2). The vast majority of patients were male. Median follow up time 16 months. The majority (57%) of patients underwent surgical management without adjuvant or neoadjuvant chemoradiotherapy. 7 patients (12%) underwent endomucosal resection. O’Pech et al reported the first case to be successfully treated with endomucosal resection in 2001 [6].
Immunohistochemical staining in this case demonstrated the tumour had a gastric immunophenotype. This tumour showed similar morphology to usual adenocarcinoma of the distal oesophagus including a papillary growth pattern. However, specialist immunohistochemical staining demonstrated the presence of adjacent gastric oxyntic type mucosa in the absence of Barrett’s epithelium, which represents a gastric variant of adenocarcinoma currently regarded as non-Barrett’s associated adenocarcinoma. Recent studies suggest that most Barrett-related carcinomas can be either gastric or intestinal type, with phenotypic stability during progression to cancer, supporting separate gastric and intestinal pathways of carcinogenesis [10]. In summary, the pathway of carcinogenesis in this case is clearly different to those adenocarcinomas associated with Barrett’s, this is also supported by the absence of intestinal metaplasia. The reporting of intestinal metaplasia amongst other case reports is highly variable, and in the majority of cases was not commented upon. Subsequently, it is difficult to draw any precise conclusions to the usual development of proximal oesophageal adenocarcinomas within EGM.

Where cases of adenocarcinomas arise in unusual locations we would recommend the use of immunohistochemical stains such as MUC-1, MUC-2, MUC-5AC, MUC-6 and CDX2, in addition to the more commonly used stains CK7 and CK20, as well as careful sampling of background mucosa to help differentiate between intestinal type and non-intestinal type adenocarcinomas. Most surgeons and endoscopists consider EGM to be a rare and benign phenomenon but this case reminds us that malignant transformation can occur within these areas.
Table 2
Summarised data of gastric adenocarcinomas of the upper oesophagus.

| Age (median (range)) | 64 (43–88) |
|----------------------|------------|
| Male:Female          | 43:5       |
| Follow up (months) (median (range)) | 16 (3–72) |

Pathology

| Well       | 13, 22% |
| Moderate   | 11, 19% |
| Poorly     | 10, 17% |
| Other      | 19, 33% |
| Unknown    | 5, 9%   |

Management

| Surgical only | 27, 47% |
| Neoadjuvant chemo and surgery | 4, 10% |
| Surgery and adjuvant chemo/rad | 3, 5% |
| Radiotherapy only | 6, 10% |
| ERM         | 7, 12%  |
| Palliative  | 1, 2%   |
| Other       | 3, 5%   |
| Unknown     | 5, 9%   |

7. Conclusion

This literature review of adenocarcinomas arising from EGM within the proximal oesophagus is the most complete to date. We discovered 55 papers reporting 58 cases of adenocarcinoma arising within EGM dating back to 1950. The management of these cases has evolved over time, but overwhelmingly the most common strategy has been oesophagectomy. More recently, small carcinomas confined to the submucosal surface have been successfully managed with endomucosal resection [11–13]. Ours is the first case to describe true gastric type adenocarcinoma arising within ectopic gastric mucosa in the proximal oesophagus. In summary, we can propose two different pathways for the pathogenesis of adenocarcinoma within EGM:

1. Induction of metaplasia-dysplasia pathway via locally secreted acid or refluxed acid leading to intestinal metaplasia and an intestinal type adenocarcinoma
2. Intrinsic development of adenocarcinoma within gastric/foveolar cells in EGM leading to gastric type adenocarcinoma

EGM although common is scarcely biopsied, whilst malignant progression is very rare, endoscopists and surgeons should be wary of the potential. The relationship between EGM and the risk for developing proximal adenocarcinomas requires much larger, population studies.

Conflicts of interest

Nil.

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Ethical approval

Not applicable.

Consent

Yes written consent obtained.

Author contribution

Ahmed Aly – conceptualisation of case report.

G. Riddiough – data collection, writing of paper.
S. Horny – writing of paper.
K. Asadi – interpretation of histology and immunohistochemistry results for paper.

Guarantor

Ahmed Aly.

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