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Case Report

Not always a distant metastasis: Can be a double malignancy too

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ABSTRACT

The incidence of double malignancy reported are between 0.3% and 4.3%. The second primary lesion is identified either simultaneously with the primary lesion (synchronous) or after a period of time (metachronous). We report a case in a 63-year-old male presenting with double malignancy with adenocarcinoma of the ascending colon and a pre-sacral Chordoma of which the latter was detected accidentally on imaging. The patient underwent surgical excision of the pre-sacral mass and an exploratory laparotomy with Right Hemicolectomy.

The increasing incidence of multiple malignant tumors is a real challenge to both clinicians and pathologists and close attention should be paid to imaging and histologic findings to avoid a misdiagnosis. In addition an early diagnosis is essential to achieve optimal treatment. We believe that the treatment modalities should be carefully tailored for each individual patient diagnosed with a double malignancy.

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1. Introduction

The presence of a second synchronous or metachronous malignancy is not uncommon. The neoplasms are either limited to a single organ or may involve multiple organs at different anatomical sites. Multiple Primary Malignancies (MPM) were first described by Billroth in 1879. The North American Association of Central Cancer Registries (NAACCR) classifies MPM into two categories: (1)- Synchronous, in which the second tumour occurs within 6 months of the primary tumour (The Surveillance Epidemiology and End Results Programme (SEER) definition is within two months) and (2)- Metachronous, in which the tumours follows in sequence, that is, more than six months apart.¹ Metachronous malignancies are more common because of the increase in the number of elderly cancer survivors as a result of improved diagnostic and treatment modalities. However the exact pathophysiology of

multiple primary tumours remains unknown.

Warren and Gates criteria² for the diagnosis of multiple primary malignancies are as follows-

1. Each tumour must be malignant and confirmed by histology.
2. Each must be geographically separate and distinct. The lesion should be separated by normal mucosa.
3. Probability of one being the metastasis of the other must be excluded.

We report a case of synchronous malignancies in a 63-year-old male diagnosed with Adenocarcinoma of the Ascending Colon and a simultaneous pre-sacral Chordoma both histologically proven.

2. Case Report

A 63-year-old South- Asian male presented with complaints of low backache and discomfort in the sitting position since 4 months. On CECT abdomen, a lobulated enhancing mass

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measuring 6.7x4.3cms was seen in the pre-sacroccygeal region causing erosion of the underlying bones. There was a short segment irregular circumferential wall thickening involving proximal ascending colon located approximately 2 cm from the caecum. On colonoscopy, the evidence of extrinsic compression of the rectum was seen along with a sigmoid colon polyp and an ascending colon growth.

Rectal EUS guided biopsy was done from the pre-sacral mass at another centre and was reported as suspicious for malignancy. Sigmoid Polypectomy was done, followed by histopathology which revealed microscopic foci suspicious for invasive adenocarcinoma in the background of adenomatous polyp with high grade dysplasia.

Ascending colon growth biopsy revealed poorly-differentiated adenocarcinoma with signet ring cell morphology. The patient was subsequently referred to this hospital.

A whole body PET – CT, showed increased FDG uptake in the ascending colon growth and pre-sacroccygeal mass, the latter interpreted as possible metastasis from the former. His laboratory parameters including the differential blood count, transaminases, alkaline phosphatase, electrolytes, BUN, creatinine and serum CEA (1ng/ml) were within normal limits.

3. Pathologic Findings

The biopsies done outside were reviewed and the patient was planned for Exploratory Laparotomy with Right Hemicolectomy with ileo-transverse anastomosis with Pre-Sacral tumour excision and sigmoid polypectomy was performed through midline vertical laparotomy incision and midline incision posteriorly. Post-operatively, the specimen was sent to the Department of Anatomic Pathology of our hospital. Grossly, the Right Hemicolectomy was measuring 32 cm in length with a polypoidal growth in the ascending colon. On serial sectioning the tumour was measured 6x4x3cms. The marginal excision of the pre-sacral mass measured 4.5x4x3 cm. On histopathologic examination the ascending colon showed a poorly differentiated adenocarcinoma with focal mucinous and signet cell areas, invading through the muscularis propria into the pericolorectal tissue with uninvolved lymph nodes. [Figure 1 A,B] Immunohistochemically, there was loss of expression of MLH1 and PMS2.

The sections from the pre-sacral mass revealed features of High grade chordoma, NOS extending into soft tissue with MF-0-1/10 hpf, and 10% of the tumour area showing necrosis.[Figure 1 C,D] Immunohistochemically, the tumour cells showed diffuse strong immunoreactivity for CK, EMA, S-100 and Brachyury.[Figure 2 A-D]

Patient is on regular follow up and clinically there is no evidence of recurrent disease till date.

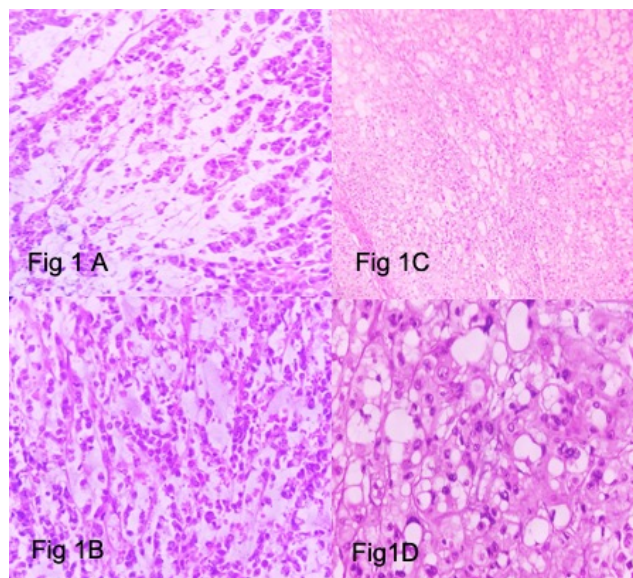


Fig. 1: A,B: Microphotograph showing Poorly differentiated Adenocarcinoma with focal mucinous and signet ring cell areas. (H & E x400); C,D: Microphotograph showing epithelioid cells with abundant clear cytoplasm giving a vacuolated appearance - High Grade Chordoma, NOS (H & E X400)

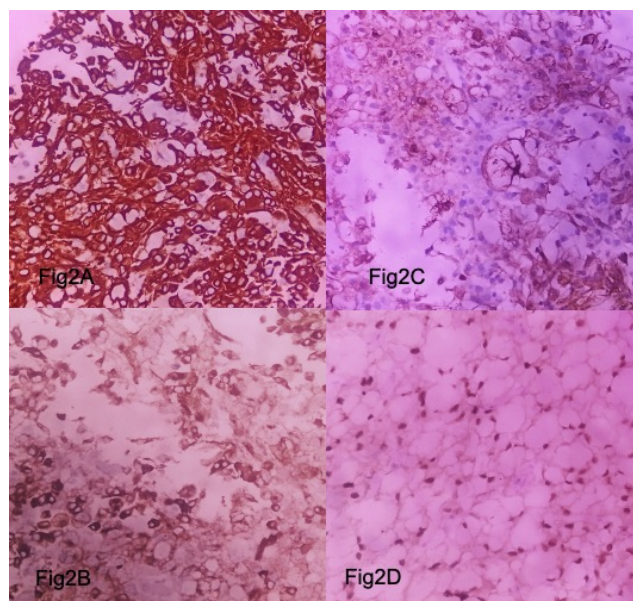


Fig. 2: A: Immunoreactivity score 4+ in neoplastic physaliferous cells (IHC stain CK X400); B: Immunoreactivity score 4+ in neoplastic physaliferous cells (IHC stain EMA X400); C: Immunoreactivity score 4+ in neoplastic physaliferous cells (IHC stain S-100 X400); D: Immunoreactivity score 4+ in neoplastic physaliferous cells (IHC stain Brachyury X400)

4. Discussion

Multiple primary tumours are a well-known and common phenomenon. Rapid advancements in diagnostics and therapeutics have contributed to a significant improvement in the survival rates of cancer patients. Operable synchronous primary malignancies can be resected in a single setting with minimal morbidity and better survival and lessening the burden on patients both psychologically and financially. Data from developing countries pertaining to synchronous and metachronous primary malignancies is very limited.

Bongers et al. concluded that, in addition to external carcinogens, an intrinsic susceptibility may influence the risk for the development of second primary tumors in patients with head and neck squamous cell carcinoma.³

Mitchell reported a patient with five primary synchronous neoplasms of the gastrointestinal tract, involving the stomach, small bowel, and colon.⁴

Synchronous primary cancers of the breast and cervix as reported by us has also been reported by Verstovsek et al.⁵ An interesting case of double malignancy-carcinoma lung and rhabdomyosarcoma of scapula-has also been reported by Masood et al.⁶ These cases illustrate the need for a thorough search for additional neoplasms when treating patients with cancer.

It is imperative that patient with a primary malignant tumors should be thoroughly, closely, and regularly followed up. Genetic counseling, risk estimation, cancer screening and chemoprevention must be emphasized.

5. Conclusion

Possibility of a synchronous or metachronous second primary malignancy should always be considered whenever a second lesion is detected at an unusual site. Histopathological evaluation of the lesion will lead to accurate diagnosis, staging, and appropriate treatment.

Appropriate cancer prevention strategy with proper emphasis on synchronous and metachronous cancer needs to be designed and incorporated into the National Cancer Control Programme as multiple primary cancers have a unique, biological behavior requiring specific diagnostic

and therapeutic intervention.

The current study emphasizes the need for a thorough and competent pre-operative biopsy evaluation in cases with multiple tumours accompanied by appropriate IHC as applicable to ensure accurate diagnosis and consequent appropriate therapeutic management.

6. Source of Funding

None.

7. Conflict of Interest

The authors declare that there is no conflict of interest.

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