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A Case of Pagets Disease of Perianal Region

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Abstract

Pagets disease is commonly associated with breast malignancy. Here we are describing about a rare presentation of pagets disease at perianal region. In our case the pagets disease was not associated with any other invasive malignancy of anal canal or rectum.

Keywords: Pagets disease, Invasive Carcinoma, Perianal pigmentation.

Introduction

Paget's disease is characterized by its presentation as dermatitis/ eczematous appearance. Patients usually have a history of itching, pigmentation and colour change. Usually, it is a subacute to chronic presentation. Paget's disease is usually associated with breast invasive carcinoma. Paget's disease diagnosis warrants a detailed investigation to look for any invasive component in the underlying tissues.

Case Report

A 74-year-old gentleman with complaints of perianal itching and pigmentation for past 10years. The size of the perianal skin pigmentation steadily increased in size. He had episodes of bleeding from the lesion on and off for past 10 days. He is a known case of hypertension and diabetes mellitus.

Under General anesthesia biopsy was taken from the perianal lesion, which showed pagets disease.



Fig. 1: preoperative picture.

Given the strong association between EMPD (Extra Mammary Paget's Disease) and underlying malignancy, unsampled colorectal adenocarcinoma was suspected. To evaluate the possibility of underlying malignancy, - colonoscopy was performed, which revealed normal study.

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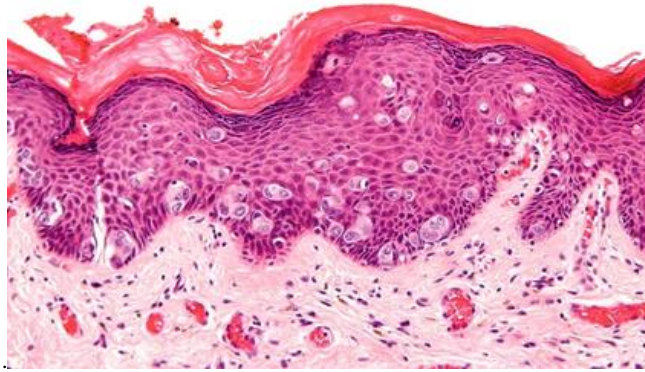


Fig. 2: Low power view of Paget cells (vacuolated cells).

The Paget cells stained positive for CK7 and CDX2 and were minimally positive for CK20 (Fig. 2).

Patient underwent wide local excision of the perianal lesion with diversion colostomy with local pedicled flap reconstruction for soft tissue defect.

Diversion loop colostomy was reversed after 4 months.

Through good rehabilitation and pelvic floor exercises patient has good anal continence.

The patient is being followed till submission of this manuscript by physical examination and anoscopy and there is no recurrence of the disease.



Fig. 3: postoperative followup picture of perianal region.

Discussion

Paget's disease of anal canal is an extremely rare condition based on rare case reports [3]. Some reports in the literature which discuss Paget's disease of the anus do indeed describe perianal Paget's disease. Given the anatomic association of the anal canal and perianal skin and the extreme rarity of anal canal lesions, these two entities can be discussed together.

EMPD is a very rare condition with variable histology amongst different cases. In some cases, Paget's cells are predominant in basal layer while in others Paget's cells are spread throughout the epidermis. Moreover, the appearance of Paget's cell is different and is described as type A (classical) and type B (signet ring like). Two different hypotheses are proposed for the development of EMPD. According to the first hypothesis, it is a neoplasm of apocrine cells occurring within skin epidermis, while the other hypothesis suggests a "pagetoid" migration of malignant cells from an underlying malignancy such as anorectal adenocarcinoma [6].

Paget's disease needs to be differentiated from other lesions with similar morphological features. Certain reactive changes such as pagetoid dyskeratosis are seen with

hemorrhoidal disease. Also, human papilloma virus (HPV) induced lesion with or without anal intraepithelial neoplasm, Bowen's disease, and melanocytic lesions can mimic Paget's disease. Histochemical markers such as mucicarmine, periodic acid-Schiff and immunohistochemical markers such as CK7, CK20, GCDFP15, CDX2, S-100, p16, melan A, and CEA can be employed to establish the diagnosis among these differentials.

This may represent a phenomenon similar to Paget's disease of breast where its association with ductal carcinoma *in situ* is relatively common [9]. One of the leading hypotheses for the pathogenesis of Paget's disease of breast is secretion by keratinocytes of epidermal heregulin- α (neuregulin I) which induces chemotaxis of Her-2 positive malignant cells to epidermis [11]. Overexpression of Her-2 is noted on the membrane and in the cytoplasm of approximately 25% and 66% of colorectal cancer cases, respectively [13] and is also reported on colorectal adenomas [15]. Similar to breast, Her-2 positive malignant cells of colorectal adenocarcinoma or tubular adenoma may migrate toward the anal mucosa or perianal epidermis due to chemotaxis in perianal EMPD.

Conclusion

We report an exceedingly rare case of anal Paget's disease. Immunohistochemistry can aid in rendering a correct diagnosis and in distinguishing primary from secondary EMPD, a distinction of important therapeutic and prognostic significance. In our case the paget's disease was not associated with any underlying invasive carcinoma survived through scopy and radiological investigation regularly till now. The hypothesis of the disease originating from the perianal skin apocrine gland might hold good in our case.

References

1. Siponen E, Hukkinen K, Heikkila P, Joensuu H, Leidenius M. Surgical treatment in Paget's disease of the breast. *Am J Surg.* 2011;200:241-246. <http://www.ncbi.nlm.nih.gov/pubmed/20678619>
2. Seetharam S, Fentiman IS. Paget's disease of the nipple. *Women's Health.* 2009;5:397-402.
3. Caliskan M, Gatti G, Sosnovskikh I, et al. Paget's disease of the breast: the experience of the European Institute of Oncology and review of the literature. *Breast Cancer Res Treat.* 2008;112:513-521. <http://www.ncbi.nlm.nih.gov/pubmed/18240020>
4. Subramanian A, Birch H, McAvinchey R, Stacey-Clear A. Paget's disease of uncertain origin. *Int Semin Surg Oncol.* 2007;4:12. <http://www.ncbi.nlm.nih.gov/pubmed/17480238>
5. Laronga C, Hasson D, Hoover S, et al. Paget's disease in the era of sentinel lymph node biopsy. *Am J Surg.* 2006;192:481-483. <http://www.ncbi.nlm.nih.gov/pubmed/16978954>
6. Marcus E. The management of Paget's disease of the breast. *Curr Treat Options Oncol.* 2004;5:153-160. <http://www.ncbi.nlm.nih.gov/pubmed/14990209>
7. Sakorafas GH, Blanchard K, Sarr MG, Farley DR. Paget's disease of the breast. *Cancer Treat Rev.* 2001;27:9-18. <http://www.ncbi.nlm.nih.gov/pubmed/11735019>
8. Bodnar M, Miller OF 3rd, Tyler W. Paget's disease of the male breast associated with intraductal carcinoma. *J Am Acad Dermatol.* 1999;40:829-831. <http://www.ncbi.nlm.nih.gov/pubmed/10321627>
9. Kollmorgen DR, Varanasi JS, Edge BS, Carson WE 3rd. Paget's disease of the breast: a 33-year experience. *J Am Coll Surg.* 1998;187:171-177. <http://www.ncbi.nlm.nih.gov/pubmed/9704964>
10. Kao GF. Mammary Paget Disease. Medscape. Last Update May 12, 2016. Available at: <http://emedicine.medscape.com/article/1101235-overview> Accessed June 21, 2016.
11. Molina GE, Khalifian S, Mull JL, Chen L, Rosman IS, Faulkner-Jones BE, Ngo KH, Demehri S, Cornelius LA, Wu PA. Topical Combination of Fluorouracil and Calcipotriene as a Palliative Therapy for Refractory Extramammary Paget Disease. *JAMA Dermatol.* 2019 May 01;155(5):599-603. [PMC free article] [PubMed]
12. Jones MA, Edwards T, Ermolovich T. Brown macule on vulva of an elderly woman. *JAAD Case Rep.* 2019 Feb;5(2):156-158. [PMC free article] [PubMed]
13. Cervantes J, Rosen A, Cho JH. Enlarging Bump on the Scrotum. *Skin Appendage Disord.* 2018 Nov;5(1):52-55. [PMC free article] [PubMed]
14. Valle L, Deig C, Wright R, High W. An advanced case of extramammary Paget disease: Safe and effective treatment in an inoperable elderly patient using extensive en face electron irradiation. *JAAD Case Rep.* 2019 Jan;5(1):72-74. [PMC free article] [PubMed]
15. Asel M, LeBoeuf NR. Extramammary Paget's Disease. *Hematol Oncol Clin North Am.* 2019 Feb;33(1):73-85. [PubMed]
16. Yao H, Xie M, Fu S, Guo J, Peng Y, Cai Z, Jiang Y, Zheng D, Wang Z. Survival analysis of patients with invasive extramammary Paget disease: implications of anatomic sites. *BMC Cancer.* 2018 Apr 10;18(1):403. [PMC free article] [PubMed]