v. 2, n. 55, 2022

International Journal of Health Science

EPIDERMOLYSIS BULLOSA AS A PREDICTOR FOR SPINOCELLULAR CARCINOMA

Anna Gabriela Figueiredo de Almeida
Faculdade de Ciências Médicas da Paraíba-FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0002-5760-2745

Ana Raissa de Melo Andrada Loureiro
Faculdade de Ciências Médicas da Paraíba-FCMPB (Afya)
João Pessoa- Paraíba
http://lattes.cnpq.br/3256829528693147

Andressa Lisboa de Carvalho Facundo
Faculdade de Medicina Nova Esperança (FAMENE)
João Pessoa- Paraíba
http://lattes.cnpq.br/5293373711880050

Izadora Barbosa Mendes
Faculdade de Ciências Médicas da Paraíba-FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0003-2151-2171

Manuelli Monteiro Meira Bastos
Faculdade de Ciências Médicas da Paraíba-FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0001-9386-0621

Mariana Cordeiro de Souza
Faculdade de Ciências Médicas da Paraíba-FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0003-3079-1379

Mayanna Meneses Xavier de Melo
Faculdade de Medicina Nova Esperança (FAMENE)
João Pessoa- Paraíba
https://orcid.org/0000-0002-9174-8190

Melissa Soares de Queiroz Rabelo Dias
Faculdade de Ciências Médicas da Paraíba- FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-002-0952-5896

Roberta Gracielle Amorim de Queiroz
Faculdade de Ciências Médicas da Paraíba- FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0002-5371-685X

Victor de França Oliveira
Residente em Cirurgia Cardiovascular do Instituto de Medicina Integral Professor Fernando Figueira- IMP
Recife- Pernambuco
https://orcid.org/0000-0001-5925-5292

Yanne Moreira Leite Loureiro
Faculdade de Ciências Médicas da Paraíba- FCMPB (Afya)
João Pessoa- Paraíba
https://orcid.org/0000-0002-6842-6484

Abstract: Epidermolysis bullosa (EB) is a group of rare heterogeneous genodermatoses characterized by mechanical fragility of epithelial tissues, blistering or prototypical erosions in mucocutaneous membranes, and impaired wound healing. Unresolved scarring results in fibrosis and often leads to associated morbidities, including cancer. Patients with EB, exposed to several complications by the continuous mechanical or pathological insult to the skin, have Cutaneous Squamous Cell Carcinoma (SCC) as the most feared of them. The aim of this study was to correlate how epidermolysis bullosa can be a predictor for squamous cell carcinoma, understanding the pathogenesis involved. It is an integrative review in the years 2017 to 2021, based on scientific articles indexed in the Virtual Health Library, using the descriptors epidermolysis bullosa, carcinoma and squamous cell separated by the Booleans "AND". The search was submitted to filters such as full text and English language, and to selection criteria, which resulted in 8 articles selected to compose the study. EB is divided into four types: simplex EB (EBS), junctional EB (JEB), dystrophic EB (DEB), subdivided into dominant DEB (DDEB) and recessive DEB (RDEB), and Kindler syndrome. Especially in the JEB and DEB variants, skin fragility leads to possible complexities, such as the alarming incidence of SCC. Unlike the SCCs that normally occur in the population due to chronic ultraviolet exposure, SCCs associated with EB tend to appear at sites of blistering, wounds and chronic scarring on the skin. The repetitive cycle of tissue damage and repair, leading to unresolved scarring, causes deterioration of cell differentiation and accumulation of carcinogenic mutations, potential for pathogenesis. SCCs can be clinically difficult to identify in patients with EB, as they resemble areas of non-malignant wounds and ulcers. In patients

with RDEB, the expression of the fibroblast gene is different from that of non-RDEB fibroblasts, and this distinction potentiates the adhesion, invasion and growth of the SCC, so the pathology is more debilitating in these patients, especially in those who have the severe generalized form. (RDEB-SG). In this type of EB, patients develop SCC at a younger age and the cumulative risk of development increases with age and is accompanied by death. As for patients with other forms of EB, as in EBS, the tendency to SCC is lower or there is no increased risk, SCC tumors that normally occur in the population due to chronic ultraviolet exposure, SCCs associated with EB usually arise in sites of formation of blisters, wounds and chronic scars on the skin. The repetitive cycle of tissue damage and repair, leading to unresolved scarring, causes deterioration of cell differentiation and accumulation of carcinogenic mutations, potential for pathogenesis. SCCs can be clinically difficult to identify in patients with EB, as they resemble areas of non-malignant wounds and ulcers. In patients with RDEB, the expression of the fibroblast gene is different from that of non-RDEB fibroblasts, and this distinction potentiates the adhesion, invasion and growth of the SCC, so the pathology is more debilitating in these patients, especially in those who have the severe generalized form. (RDEB-SG). In this type of EB, patients develop SCC at a younger age and the cumulative risk of development increases with age and is accompanied by death. For patients with other forms of EB, as in EBS, the tendency to SCC is lower or there is no increased risk, the tumors.

Keywords: Squamous cell carcinoma, Epidermolysis bullosa, Genodermatoses.

REFERENCES

Cutaneous Squamous Cell Carcinoma in Epidermolysis Bullosa: a 28-year Retrospective Study. Robertson, S. J. et al. Cutaneous Squamous Cell Carcinoma in Epidermolysis Bullosa: a 28-year Retrospective Study. Acta Derm Venereol. 2021.

Epidermolysis Bullosa-Associated Squamous Cell Carcinoma: From Pathogenesis to Therapeutic Perspectives. Condorelli, A.G.; Dellambra, E.; Logli, E.; Zambruno, G.; Castiglia, D. Epidermolysis Bullosa-Associated Squamous Cell Carcinoma: From Pathogenesis to Therapeutic Perspectives. Int. J. Mol. Sci. 2019.

Epidemiology and Outcome of Squamous Cell Carcinoma in Epidermolysis Bullosa in Australia and New Zealand. Kim, M., Li, M., Intong-Wheeler, L. R., Tran, K., Marucci, D., & Murrell, D. F. Epidemiology and Outcome of Squamous Cell Carcinoma in Epidermolysis Bullosa in Australia and New Zealand. Acta Dermato-Venereologica. 2017.

Extracellular Vesicles as Biomarkers for the Detection of a Tumor Marker Gene in Epidermolysis Bullosa-Associated Squamous Cell Carcinoma. Sun, Y. et al. Extracellular Vesicles as Biomarkers for the Detection of a Tumor Marker Gene in Epidermolysis Bullosa-Associated Squamous Cell Carcinoma. J Invest Dermatol. 2018.

Genetic Profiles of Squamous Cell Carcinomas Associated with Recessive Dystrophic Epidermolysis Bullosa Unveil NOTCH and TP53 Mutations and an Increased MYC Expression. Sans-DeSanNicolas, L. et al. Genetic Profiles of Squamous Cell Carcinomas Associated with Recessive Dystrophic Epidermolysis Bullosa Unveil NOTCH and TP53 Mutations and an Increased MYC Expression. J Invest Dermatol. 2018.

Impaired Wound Healing, Fibrosis, and Cancer: The Paradigm of Recessive Dystrophic Epidermolysis Bullosa. Tartaglia, G.; Cao Q.; Padron, Z. M.; South, A. P. Impaired Wound Healing, Fibrosis, and Cancer: The Paradigm of Recessive Dystrophic Epidermolysis Bullosa. Int J Mol Sci. 2021.

Multiple Skin Squamous Cell Carcinomas in Junctional Epidermolysis Bullosa Due to Altered Laminin-332 Function. Furtugno, P. et al. Multiple Skin Squamous Cell Carcinomas in Junctional Epidermolysis Bullosa Due to Altered Laminin-332 Function. Int J Mol Sci. 2020.

Reprogramming and Differentiation of Cutaneous Squamous Cell Carcinoma Cells in Recessive Dystrophic Epidermolysis Bullosa. Rami, A.; Łaczmański, Ł.; Jacków-Nowicka, J.; Jacków, J. Reprogramming and Differentiation of Cutaneous Squamous Cell Carcinoma Cells in Recessive Dystrophic Epidermolysis Bullosa. Int J Mol Sci. 2020.