

Aggressive Digital Papillary Adenocarcinoma—Case Series and Meta-Analysis of a Rare Hand Tumor

Shaun D. Mendenhall, MD, Jennifer L. Koechle, MPH, CCRP, Steven J. Verhulst, PhD and Michael W. Neumeister, MD

Institute for Plastic Surgery, Southern Illinois University School of Medicine, Springfield, IL

Objectives

Aggressive digital papillary adenocarcinoma (ADPA) is a rare skin adnexal tumor with predilection for the hand. Because the rarity of this tumor, published data include only case reports or small case series, making it exceptionally difficult to define an appropriate treatment algorithm. We aim to clarify predisposition for the disease and the optimal initial treatment.

Methods

We searched the Medline database for case reports and descriptions of ADPA. We included reports of individual variables and excluded reports without case-specific information. We included articles that reported data that had been averaged and compared the results of this meta-analysis with the results of other published case series.

67 articles



217 cases



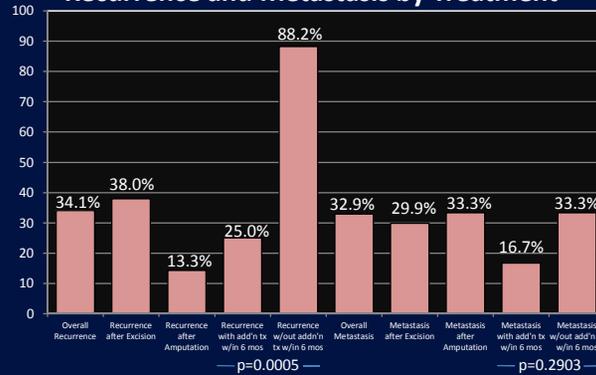
174 cases included

Results

217 cases were mentioned in the literature, 174 of which were included in this aggregate data analysis (including 3 of our own patients). Similar to prior published data, mean patient age was 47 (range 11-85), and 82% were male. Average tumor size was 2 cm and 82% were on the upper extremity. Duration of lesion until diagnosis was 4.6 years, and 5.2% had a history of trauma. Common treatment was excision (88%) vs. amputation (11%). The overall recurrence rate was 34%, and overall metastasis rate was 33%. Recurrence rate by initial treatment was 38% for excision and 14% for amputation. Metastasis rate was 30% for excision and 33% for amputation. At last follow up, 80% of patients were alive and well, with another 11% living with the disease, 6% dying of the disease, and 3% dying of unrelated causes. We report an average follow up of 45.6 months.



Recurrence and Metastasis by Treatment

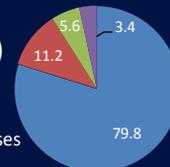


ADPA Case Means

	All	< 25 years old
Age (years)	47.2	17.4
Upper extremity (%)	81.5	27.3
Duration (years)	4.6	1.2
Size (cm)	2.0	2.2
Recurrence (%)	34.1	42.9
Follow up (years)	3.8	4.8

Distribution of Outcomes (%)

■ Alive & well
■ Alive w/ disease
■ Died of disease
■ Died, unrelated causes



Discussion

Our data supports that re-excision or amputation within 6 months of initial treatment yields a significantly lower recurrence rate.¹ We found that if re-excision or amputation was performed within 6 months of initial treatment, the recurrence rate after 6 months drops to 25.0%, compared to 88.2% if subsequent treatment is not performed within 6 months (p=0.0005).

Also, the overall metastatic rate of our patients was 33%, which was higher than previously reported rates of 14% and 26%.^{1,2} Comparing patients with subsequent treatment within 6 months also showed a decrease in metastatic rate from 33% in those without additional treatment within 6 months to 17% who had subsequent treatment within 6 months, but this finding was not statistically significant (p=0.2903).

Interestingly, in 11 cases in patients less than 25 years old, 73% of tumors occurred on the lower extremity, and 55% of tumors appeared on females. Half of these younger patients experienced recurrence, with average recurrence time of 25 months. The disease metastasized in 43%, with average metastasis time of 83 months.

1. Duke W, Sherrod TT, Lupton G. Aggressive digital papillary adenocarcinoma (aggressive digital papillary carcinoma revisited). *Am J Surg Pathol* 2000;24:775-84.
 2. Suchak R, Wang WL, Prieto V, et al. Cutaneous Digital Papillary Adenocarcinoma. A Clinicopathologic Study of 31 Cases of a Rare Neoplasm With New Observations. *Am J Surg Pathol* 2012; 36(12): 1883-91.

Conclusions

To our knowledge, the current report is the only meta-analysis of published ADPA cases. Our aggregate data analysis supported previously described demographics, except for ADPA cases in patients younger than 25. However, it does not give clear guidance on treatment choice. Metastasis rate was somewhat less in patients who had amputations, but this was not statistically significant. Due to implications of amputation on form and function, wide local excision may offer similar benefits without the detriments of the loss of a digit. A second surgical procedure within 6 months was shown to significantly reduce recurrence. We concur with the published information on the importance of including ADPA in the differential diagnosis when evaluating tumors of the hand.